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با محوریت سندروم  
کودکان ایران

ششمین کنگره  
بین المللی  
انجمن  
نفرولوژی



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# ABSTRACT BOOK

## Oral Presentation

### *Article No 18102*

#### **Pathogenesis of edema in nephrotic-nephritic syndrome, new edema**

Hasan Otoukesh

Nephritic syndrome is a collection of signs (syndrome) associated with disorders affecting the kidneys more specifically glomerular structure, characterized by: having small pores in the podocytes, large enough to permit proteinuria and hematuria (RBC, RBC cast), leukocyturia (WBC cast). Edema is shown in the nephritic syndrome by accumulation of salt and water in the vessels with overload manifestations. Salt restriction during the acute phase to control edema and volume related HTN with diuretic and antihypertensive drugs are recommended.

Nephrotic syndrome is a syndrome with hypoalbuminemia, hyperlipidemia, edema and massive proteinuria. Edema occurrence in nephrotic syndrome is different from nephritic syndrome. Edema is seen in these patients by accumulation of salt and water in extravascular space. Salt restriction is advised for the prevention and treatment of edema, a very low salt diet is only necessary in cases of massive edema (33mg/kg/day). Fluid restriction is recommended in moderate to severe hyponatremia (PNa<125 meq/l), continue weight gain against salt restriction.

### *Article No 18103*

#### **Simplified algorithm for evaluation of asymptomatic persistent isolated proteinuria**

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**Background** - Proteinuria is a common laboratory finding among children and adolescents. It can be identified as either a transient or a persistent finding and can represent a benign condition or a serious disease.

**Methods** – Pertinent medical literature for asymptomatic proteinuria in children and adolescents published in English was searched between January 1980 and May 2017 using PubMed, MEDLINE, EMBASE, and Google Scholar research databases.

**Results** - Proteinuria has numerous etiologies, but nonspecific symptoms. Random spot urine protein-to creatinine (Pr/Cr) ratio is widely used to detect proteinuria. In adults and children over two years of age, spot urine Pr/Cr < than 0.3 is considered normal. In children aged six months to two years, the upper limit of normal is 0.5. Orthostatic proteinuria is defined as urine Pr/Cr>0.3 detected during the day time activity but <0.3 on the first morning urine specimen. Urine Pr/Cr above 3.0 is found in patients with nephrotic syndrome.

Orthostatic proteinuria is a frequent cause of proteinuria in asymptomatic children and adolescents, which require no specific therapy except for health maintenance follow-up. Pediatric nephrologist referral is indicated when the proteinuria is constant and persists over 6 months or is associated with hematuria, hypertension or renal dysfunction.

**Conclusion** - we provide a simplified diagnostic algorithm for evaluation of proteinuria in children who appear well and in whom proteinuria is incidentally discovered during a routine examination.

**Keywords:** Adolescent, Algorithms, Adolescent, Child, Proteinuria

### *Article No 18104*

## **Systematic Review of Efficacy and Safety of Ofatumumab in Children with Difficult-to-Treat Nephrotic Syndrome**

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**Objectives:** Different types of studies have been done so far on drugs efficacy and safety in children with refractory nephrotic syndrome. Ofatumumab might be an effective treatment for this syndrome; however, the long-term effects and cost-effectiveness of Ofatumumab treatment have not been comprehensively assessed. This study aims to do a systematic review on the efficacy and safety of Ofatumumab in children with difficult-to-treat nephrotic syndrome.

**Material and Methods:** An electronic literature search was conducted to identify appropriate studies. The search term was: ("nephrotic syndrome" or "minimal change disease" or "focal segmental glomerulosclerosis" or membranous) and ("Ofatumumab" or "CD20" or "Arzerra" or "HuMax-CD20"). We included all studies related to using Ofatumumab in children with difficult-to-treat nephrotic syndrome. Two independent reviewers extracted data from the articles according to the selection criteria.

**Results:** Eligible studies were included in this systematic review. The literature search and reference mining yielded 77 potential relevant articles. We removed 32 articles because of duplication. Also, 26 references were excluded after reviewing the titles and abstracts because they were books, book sections, review papers and therefore not relevant. Then, we reviewed full-text of selected articles and removed 14 other studies because the topics were not relevant to the subject. At last, 5 studies were included in the systematic review. The metric considered to assess the efficacy of Ofatumumab in children with nephrotic syndrome in most of the studies was complete remission rate.

**Conclusions:** Our systematic review shows that Ofatumumab may be effective in the treatment of refractory nephrotic syndrome in children, and it can reduce the use of steroid and immunosuppressants. However, acknowledging the limitations of the study due to the size and nature of the studies included, further large randomized trials are suggested.

**Keywords:** Ofatumumab, Nephrotic syndrome, Children, Treatment

### *Article No 18105*

## **Relapse and steroid resistance in children with nephrotic syndrome Comparison of 8 weeks and 12 weeks steroid therapy protocol**

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**Background :** nephrotic syndrome is one of the most common pure renal diseases that a pediatric nephrologist encounters in his /her day to day practice. Initial treatment of primary nephrotic syndrome in children with corticosteroid is a standard of therapy since decades ago, but duration and protocol of steroid treatment is still challenging . in this study we compared recurrence rate in two groups of children with nephrotic syndrome with 8 weeks and 12 weeks course of steroid treatment.

Materials and methods:

**Method:**This non randomized clinical trial was performed on 68 children with primary nephrotic syndrome referred to pediatric nephrology clinic. Patients were divided into two groups, group1 (34 patients )received eight weeks of oral prednisolone ( four weeks 2mg/kg/day every day divided to three doses and four weeks 1.5 mg/kg single dose alternate day) and group 2 ( 34 patients) received twelve weeks prednisolone (six weeks 2mg/kg/day every day divided to three doses and six weeks 1.5 mg/kg single dose alternate day), after one year follow up rate of relapse, steroid dependence and steroid resistance was calculated for each group.

**Results:** In group 1, mean age of patients was  $4 \pm 1.67$  year, 64.7% (22 patients) were male and 35.3% (12 patients) were female. In group 2 mean age was  $4.09 \pm 1.97$  year, 70.6% (24 patients) were male and 29.4% (10 patients) were female. Difference of age and sex distribution was not significant. In group 1 ( eight weeks steroid) rate of frequent relapses, steroid dependence and steroid resistance was 26.5%, 17.6% and 8.8% accordingly. In group 2 (twelve weeks steroid) rate of frequent relapses,steroid dependence and steroid resistance was 11.8%, 8.8% and 5.9% accordingly. Group 1 patients had 47.1% rate of remission and occasional relapse and group 2 had 73.5% of remission and occasional relapse . in comparison rates of frequent relapses, steroid dependence and steroid resistance were significantly lower in group 2 than group 1 ( P value 0.026).

**Conclusion:** twelve weeks steroid treatment protocol is associated with significant lower rate of frequent relapses, steroid dependence and steroid resistance compared to eight weeks steroid protocol, although increased risk of prolonged steroid treatment should be considered in this protocol of treatment of nephrotic syndrome.

### Article No 18106

#### Comparison of carotid intima media thickness in children with nephritic syndrome Vs healthy children

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**Introduction:** Nephrotic syndrome is one of the most common glomerulopathy within children and dyslipidemia as well as abnormal fat metabolism even during remission period can produce endothelial dysfunction and early atherosclerosis and promote cardiovascular disease (CVD). Carotid intima medial thickness (IMT) is well correlated with atherosclerosis and the aim of this study is to evaluate IMT as a risk factor of CVD in children.

**Methods:** As case-control study, 30 children with nephrotic syndrome aged between 2 and 12 years old, mean age 6.66 years with at least 2 years involvement enrolled the study. Thirty healthy children with mean age 9.93 year old selected as control group. All subjects on both groups underwent carotid ultrasound by one device and by one expert specialized operator. Data recorded and compared the results using chi square test and Fisher's exact test between two groups.

**Results:** The results showed that carotid intimal thickness in patients and healthy groups were 0.46 mm and 0.43 mm, respectively ( $p < 0.005$ ). Age, sex and BMI has no significant effect on IMT.

**Conclusion:** Long standing illness and steroid medication in children with nephrotic syndrome make them susceptible to atherosclerosis and early development of cardiovascular attack. Ultrasonographic ITM assay is a safe and non-invasive method to detect vulnerable patients.

**Key words:** Nephrotic syndrome, Carotid intima medial thickness, Atherosclerosis

### *Article No 18107*

#### **The efficacy and tolerability of rituximab in children with steroid and cyclosporine-resistant and steroid and cyclosporine-dependent pediatric nephrotic syndrome**

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**Background:** A few clinical trial-based evidences could show the effectiveness of rituximab in relieving manifestations of nephrotic syndrome especially in affected children. The present study aimed to assess the therapeutic effectiveness of Rituximab in steroid and cyclosporine-resistant and steroid and cyclosporine-dependent pediatric nephrotic syndrome.

**Methods:** This prospective interventional study was performed on 42 children aged lower than 18 years with steroid and cyclosporine-resistant or steroid and cyclosporine-dependent pediatric nephrotic syndrome who were candidate for administrating rituximab. All patients received intravenous rituximab ( $375\text{mg}/\text{m}^2$ ) per week for 4 weeks. For assessment of renal functional status, dipstick urinalysis was scheduled weekly for up to four weeks and then once a month to three months.

**Results:** Regarding underlying pathological pattern, 73.8% had FSGS, 21.4% had MCNS and 4.8% suffered MGN. Overall, 57.1% of children suffered steroid and cyclosporine-resistant nephrotic syndrome, 16.7% had drug late-resistant nephrotic syndrome and 26.2% had steroid and cyclosporine-dependent pattern of disease. Regarding response to treatment, 61.9% responded completely to treatment with Rituximab, 7.1% had partial response, 4.8% had clinical response and others remained with no significant response to this drug. Drug-related side effects were found only in 4 patients as leukopenia in two children, alopecia in one children and eosinophilia in another one. In those children who suffered steroid and cyclosporine-resistant disease, complete, partial, and clinical response was revealed in 37.5%, 12.5%, and 4.2% respectively, while in the group with late resistant pattern was 87.5%, 0%, and 14.3%, and in those with steroid and cyclosporine-dependent pediatric nephrotic syndrome was 100%, 0% and 0% respectively indicating significantly higher complete response in those with drug-dependent pattern ( $p = 0.006$ ). The number of relapses was reduced after receiving rituximab when compared to before treatment. The response rate to Rituximab did not dependent to gender, age, or disease pathology.

**Conclusion:** Rituximab is effective for children with nephrotic syndrome with high efficacy and well tolerability especially in those with steroid and cyclosporine-dependent pediatric nephrotic syndrome

### *Article No 18108*

#### **Effect of Levamisole in Steroid-Dependent Nephrotic Syndrome**

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**Introduction.** Childhood idiopathic nephrotic syndrome is characterized by frequent relapsing courses or steroid dependency.

Levamisole is a popular drug for treatment of these patients. The purpose of this study was to evaluate levamisole in children with steroid-dependent nephrotic syndrome.

**Materials and Methods.** We retrospectively studied 304 children with a diagnosis of steroid-dependent nephrotic syndrome or frequently relapsing nephrotic syndrome. The mean age at the time of diagnosis was 4.84 years.

Following induction of complete remission with steroid therapy based on the International Study of Kidney Disease in Children's protocol and when they were taking alternative days of steroid, 2.5 mg/kg of levamisole was administered.

**Results.** The steroid dose was significantly decreased (mean reduction of  $0.39 \pm 0.46$  g to  $0.33 \pm 0.38$  g) after treatment with levamisole ( $P < .001$ ). The number of relapses also significantly decreased (mean reduction of  $0.92 \pm 0.98$  episodes to  $1.07 \pm 1.20$  relapses per year;  $P < .001$ ). The 14.5-month administration of levamisole had a sensitivity of 67.5% and a specificity of 71.9% to reach a dose reduction of more than 50% in steroid therapy. The duration of levamisole treatment was associated with more than 50% reduction in the number of relapses ( $P < .001$ ). A 14.5-month treatment with levamisole had a sensitivity of 62.3% and a specificity of 63.6% to reach a relapse reduction of more than 50%.

**Conclusions.** Levamisole appears to be effective in prolonging the duration of remission and decreasing the steroid dose in children with steroid-dependent nephrotic syndrome.

### Article No 18109

## Evaluation of children with steroid resistant NS showing pathologic finding of FSGS after renal transplantation in IRAN education and treatment centers (1998-2018)

Khadije Ghasemi

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**Background and Objectives :** Steroid resistant NS with pathologic features of idiopathic or gene mutation-associated FSGS is a common cause of End Stage Renal Disease (ESRD) in children, which may lead to kidney transplantation .

Relapse of the disease in the transplanted kidney, regardless of the medical and nonmedical preventive strategies before and after the transplantation, can result in renal dysfunction.

Since there was not comprehensive information on post-transplant FSGS patients, we decided to collect data by our nephrologist colleagues' cooperation in order to plan better preventive and treatment strategies in the future.

**Methods :** A questionnaire was designed and filled by the nephrologists from different centers order to collect data on the number of transplanted FSGS patients, transplantation date, number of relapsed cases, date of relapse, pre- and post-transplantation immunosuppressive medications, pre-transplantation medical and surgical interventions, and treatment response after relapse.

**Results :** From 82 pediatric FSGS patients who underwent kidney transplantation during the years 1998-2018, 23 cases of relapse were observed (10 cases from deceased donors and 5 from preemptive or living-related donors). The time of relapse was 1 week-1 month after transplantation in 7 questionnaire, 1 month-1 year in 8, and after 1 year in the remaining. The medications used at the time of relapse were rituximab, Plasma Exchange (PE), as well as angiotensin receptor blockers (ARB), Angiotensin converting enzyme inhibitors (ACEI) ( 6 centers), methylprednisolon pulse (5 centers) and immunosuppressive drugs-which were almost the same in all the centers-.

Genetic studies had only been done in two centers-mainly during research projects- and there was no difference in the effective medication protocol between the genetic mutation associated and idiopathic FSGS patients.

In two centers, Plasma Exchange (PE), rituximab, and IVIG were administered before transplantation. Graft delayed function was seen in 9 cases and didn't occur in 7.

The pre- and post-transplantation immunosuppressive medications were corticosteroids , mycophenolate mofetil (MMF), and tacrolimus in all the centers. IL2RBs (3 centers), anti-thymoglobulin (2 centers) and cyclosporine (6 centers) were also used with the former drugs (with or without MMF).

Diagnosis of relapse was made by kidney biopsy in 11 ques ( T-number of questionnaires filled:-14). The rate of relapse was from <10% to >50% in different centers. Some cases of relapse didn't have any response to the medical interventions. The least relapse rate was seen in 2 centers (T=40, n=9) with deceased donor and administration of rituximab and PE before transplantation.

**Conclusion :** Identification of genetic mutation associated Vs idiopathic FSGS, as well as high risk patients and choosing the best preventive and treatment protocols based on the above information can help decrease the rate of relapse in these patients.

**Key Words :** Nephrotic Syndrome , FSGS , Kidney transplantation

### Article No 18110

#### Thyroid Function in Idiopathic Nephrotic Syndrome

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**Introduction:** Nephrotic syndrome is a common glomerular disease in children. It's main feature is loss of albumin and high molecular weight proteins such as thyroxin-binding globulin and thyroid hormones. Therefore subclinical hypothyroidism is proposed with increasing in TSH and normal free thyroxin and free triiodothyronine, but some nephrotic patients reveal low/normal free fractions and overt hypothyroidism. As hypothyroidism affect steroid-response and prognosis, thyroxin replacement therapy is wise until stable thyroid status is achieved.

**Method:** This is a cross-sectional study included nephrotic patients admitted and the out-patients in clinic of Qods hospital from 1395 to 1396. There were 49 children in active and 24 in remission phase. We assessed thyroid function (TSH, free T3, free T4, total T3, total T4, and anti-TPO) for the patients and for 74 healthy children matched in gender and age.

**Result:** Thirty six (73.5%) of patients in active phase had abnormal total thyroxin with positive correlation with Upr/Ucr ratio ( $p < 0.001$ ). TSH level was elevated in active phase, with positive correlation with Upr/Ucr ratio ( $p < 0.001$ ). Thirty four percent of patients needed thyroxin replacement therapy for a short time.

**Conclusion:** Due to prevalence of hypothyroidism in our nephrotic patients, we recommend routine thyroid screening and early replacement therapy. A multi-central is also proposed for further confirmation.

### Article No 18111

#### The correlation between attention deficit hyperactivity disorder and steroid-dependent nephrotic syndrome

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**Introduction:** Nephrotic syndrome (NS) is characterized by nephritic-range proteinuria and the triad of clinical findings associated with large urinary losses of protein, hypoalbuminemia, edema and hyperlipidemia. More than 80% of children below 13 years of age with primary NS have steroid-responsive forms. There is no identifiable cause of attention-deficit hyperactivity disorder (ADHD). It is likely that the symptoms of ADHD represent a final common pathway of diverse causes, including genetic, organic and environmental etiologies.

**Methods:** This case-control study was performed on 130 children aged between 5 and 13 years who were followed-up for two years. Sixty-five children with steroid-dependent nephrotic syndrome (SDNS) as the case group and 65 healthy children as the control group were included in the study. Patients with minimal change NS were treated with prednisolone for at least six months. Conner's Parent Rating Scale - 48 (CPRS-48) was completed by the parents and the children were identified with any form of ADHD. Then, children were referred to an expert psychiatrist. The collected data were analyzed with SPSS software.

**Results:** The result showed that there was no significant relationship between different types of ADHD in both groups.

**Conclusion:** Based on current study, one may conclude that there are no significant differences between prevalence of ADHD in children with SDNS and the control group.

**Key Words:** Attention Deficit Hyperactivity Disorder, Steroid-Dependent, Nephrotic Syndrome, Children

### *Article No 18112*

#### **Thromboembolic complication in children with nephrotic syndrome**

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thromboembolism is a recognized complication in children with nephrotic syndrome, although uncommon, may be associated with severe and life threatening outcomes. In this study we retrospectively reviewed cases of children with nephrotic syndrome with clinical thromboembolism.

**Methods and materials:** In this observational study we reviewed cases of 70 children with primary nephrotic syndrome referred to pediatric nephrology clinic for manifestations of overt thromboembolism.

**Results :** There were 48 male and 22 females ( male: female ratio 2.18) with mean age of  $4.5 \pm 2.45$  years . only two male patients had presented with overt thromboembolism , one 6 year old patient with hematuria and pleuritic chest pain whom thrombosis of renal vein , inferior vena cava and pulmonary emboli was diagnosed for him and another 5 year old boy with severe headache and vomiting whose brain MRV revealed dural sinus venous thrombosis. Both patients after anticoagulation therapy fully recovered and are under follow up for their nephrotic syndrome.

In this article we study prevalence of overt thromboembolism in our pediatric children with nephrotic syndrome and describe clinical manifestations and clinical outcome of these patients with a short review of thrombotic events in nephrotic syndrome.

**Conclusion:** Clinical consideration of early signs and symptoms of thromboembolism in children with nephrotic syndrome may be important in timed diagnosis and management to prevent mortality and morbidity in these group of patients.

### *Article No 18113*

#### **Outcome of children with isolated microscopic hematuria without renal biopsy**

Abolhassan Seyedzadeh , Mohammad-Reza Tohidi , Rahimpour Amiry, Mohammad-Saleh Seyedzadeh, Sara Hookary

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**Introduction:** Hematuria (presence of  $>5$  RBCs/HPF) may be a transient outcome or indicator of significant renal disorder in children. Children with neither symptoms of a disease nor a physical abnormality who have microscopic hematuria should be placed in the category of isolated microscopic hematuria (IMH).

**Objectives:** The aim of this study was to evaluate the course of IMH. **Patients and Methods:** This investigation is an observational study of 124 patients referred to pediatric nephrology clinic from 2002-2012 with IMH.

**Results:** In this study, 124 patients, 40 (32.3%) female and 84 (67.8%) male were evaluated. The mean age was  $5.6 \pm 2.4$  years. The mean follow-up time was  $14.3 \pm 14.4$  months. This mean for 45.2% of the patients, was less than 6 months and for 4% of the patients, it was more than 4 years. The reasons for discovering hematuria were; 66.1% after routine evaluation, 21.8% due to positive family history and 12.1% after urinary tract infection (UTI). In this study, all the laboratory tests and kidney function were normal, except for the presence of microscopic hematuria.

**Conclusion:** It was concluded that IMH without renal failure, hypertension (HTN) and proteinuria is a benign condition with no need for kidney biopsy.

**Keywords:** Hematuria, Proteinuria, Biopsy, Microscopic

### Article No 18114

#### Mutation analysis of the *CTNS* gene in Iranian patients with infantile nephropathic cystinosis

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Nephropathic cystinosis is an inherited lysosomal transport disorder caused by mutations in the *CTNS* gene that encodes for a lysosomal membrane transporter, cystinosin. Dysfunction in this protein leads to cystine accumulation in the cells of different organs. The accumulation of cystine in the kidneys becomes apparent with renal tubular Fanconi syndrome between 6 and 12 months of age and leads to renal failure in the first decade of life. The aim of this study was to analyze the *CTNS* mutations in 20 Iranian patients, from 20 unrelated families, all of whom were afflicted with infantile nephropathic cystinosis. In these patients, seven different mutant alleles were found, including two new mutations, c.517T>C; p.Y173H and c.492\_515del, that have not been previously reported. In addition, we observed that c.681G>A, the common Middle Eastern mutation, was the most common mutation in our patients. Moreover, a new minisatellite or variable number of tandem repeat marker (KX499495) was identified at the *CTNS* gene. Seven different alleles were found for this marker, and its allele frequency and heterozygosity degree were calculated in cystinosis patients and healthy individuals.

### Article No 18115

#### Association of Endothelin-1 rs5370 G>T gene polymorphism with the risk of nephrotic syndrome in children

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**Background:** Primary nephrotic syndrome (NS) is a common kidney disease in children.

**Objectives:** The present study was aimed to investigate whether rs5370 G>T (lys198Asn) genetic variant of endothelin-1 (ET-1) is involved in the susceptibility to NS.

**Patients and Methods:** This case-control study was performed on 138 patients with NS and 150 healthy children. Genomic DNA was extracted from whole blood using salting out method. Polymorphism of the ET-1 rs5370 G>T (lys198Asn) polymorphism detected by T-ARMS-PCR as well as PCR-RFLP method.

**Results:** The results showed that the genotype and allelic frequencies of the ET-1 rs5370 G>T variant were not significantly different between cases and controls. Furthermore, subgroup analysis showed that

rs5370 G>T variant was not associated with gender of patients. In NS patients the genotype was not associated with cholesterol, triglyceride, total protein and albumin levels.

**Conclusions:** In conclusion, our findings indicate that ET-1 rs5370 G>T is not associated with NS.

Further studies with larger sample sizes and different ethnicities are required to validate our findings.

### Article No 18116

#### TRPC6 mutational analysis in Iranian children with focal segmental glomerulosclerosis

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**Introduction:** Focal segmental glomerulosclerosis (FSGS) is a type of nephrotic syndrome which identified by edema, proteinuria, hypoalbuminemia and hyperlipidemia. TRPC6 is one of the several genes which can cause FSGS. The main aim of this study is to analyze TRPC6 in Iranian children with FSGS and evaluation the clinical features of patients with and without TRPC6 variants.

**Materials and Methods:** twenty six patients under 16 year old were recruited. Exons 2 and 13 of TRPC6 gene were analyzed by polymerase chain reaction (PCR) amplification and sequencing.

**Results:** The mean age of patients when sampling has been done was  $9.26 \pm 3.19$  years (range: 2-15 years). 16 patients (61.5 %) were male and male-female ratio was 1.35:1. TRPC6 variants were identified in 4 patients (15.4 %). We identified 3 missense nonsynonymous mutation (C121S, D130V, G162R) and one missense mutation (c.333 C>T) without causing amino acid substitution (I111I). All variants were novel and in-silico analysis predicted D130V and G162R as pathogenic. There was no significant difference in following variables between patients with and without mutations: Age at disease onset, gender, hypertension, hematuria, serum creatinine and albumin, rate of progression to CRF and ESRD, consanguinity, response to steroid, resistance to cyclosporine A and cyclophosphamide.

**Conclusions:** In this study we evaluated exons 2 and 13 of TRPC6 gene in Iranian children with FSGS. We identified four novel TRPC6 variants which two variants (D130V, G162R) by in-silico analyzing predicted as pathogenic. TRPC6 can be useful in Iranian children with FSGS for genetic screening.

### Article No 18117

#### Evaluation of mutant allele frequency of NPHS2 gene in patients involved infantile nephritic syndrome

Hassan Otoukesh, Farahnaz kamyab, Reza Taghi pour



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چکیده

**مقدمه:** موتاسیون در *NPHS2* مسئول 39 تا 48 درصد موارد فامیلی و 10 تا 28 درصد موارد اسپورادیک می باشد. این موتاسیون در بیمارانی با سندرم نفروتیک مادرزادی دیده می شود. سندرم نفروتیک فرمی شایع از بیماری کلیه در کودکان است که با ادم، پروتئینوری ماسیو، هیپوآلبومینمی و هیپرلیپیدمی مشخص می شود. اگر شروع زودرس در سال اول زندگی داشته باشد سریعاً به سمت نارسایی مزمن کلیه پیش می رود. ژن معیوب *NPHS2* شایعترین علت ژنتیکی سندرم نفروتیک در سال اول زندگی است که توارث اتوزوم مغلوب دارد و با توجه به شیوع ازدواج فامیلی در کشور بر آن شدیداً تا فراوانی ژن معیوب *NPHS2* را در موارد سندرم نفروتیک با شروع زودرس در شیرخوارگی مورد بررسی قرار دهیم.

ابزار و روش ها: مطالعه انجام شده از نوع مطالعه بررسی بیماران (CASE-SERIES) می باشد که بصورت گذشته نگر انجام شد. جمعیت مورد مطالعه این پژوهش شامل کودکان مبتلا به سندرم نفروتیک مادرزادی غیر سندرمیک بود که در بخش کلیه بیمارستان حضرت علی اصغر (ع) و رسول اکرم (ص) بستری و تحت درمان قرار گرفته بودند مدت زمان بررسی از ابتدای فروردین ماه 1389 تا ابتدای فروردین 1390 به مدت 1 سال بود. با استفاده از کیت *Diatom DNA prep* از نمونه های خون، *DNA* استخراج شد. اگزون های ژن *Podocin* به کمک روش *PCR* تکثیر گردید. نتایج: در مطالعه انجام شده بر روی 9 بیمار مبتلا به سندرم نفروتیک مادرزادی، میانگین سن 5 / 3 ± 94 / 592 ماه (حداقل 1 = و حداکثر 12 =) میانگین وزن 7027 / 3095 ± 78 / 875 گرم (حداقل 3500 = و حداکثر 13000 =) بود 6 / 55 درصد پسر و 4 / 44 درصد دختر بودند. میانگین آلبومین 1 / 0 ± 97 / 345 *mg/dl* (حداقل 1 / 5 = و حداکثر 2 / 5 =) میانگین کلسترول 430 *mg/dl* / 239 ± 13 / 002 حداقل = 268 و حداکثر ( 980 =) بود. نوع ناهنجاری ژنتیک در 1 / 11 درصد *G deletion*، 1 / 11 درصد *Splicing problem (IVS7+7 A>G)*، 2 / 22 درصد *Splicing problem (IVS7+27G>A)* و 3 / 33 درصد *Mutation NPHS2* بود؛ و شایعترین موتاسیون *rs1410592* بود که یک موتاسیون نهفته در آخرین اگزون می باشد. ویژگی های دموگرافیک شامل سن، جنس بین دو گروه اختلاف معنی دار نداشت. و تمامی بیماران دچار سندرم نفروتیک مقاوم به درمان بودند نتیجه گیری: در مجموع موتاسیون *NPHS2* در بین بیماران ایرانی مبتلا به سندرم نفروتیک شیرخوارگی غیر سندرمیک شایع بوده و با درصد های جهانی هم خوانی دارد و لذا استفاده از روش های آنالیز ژنی برای تشخیص این موتاسیون در تشخیص زودرس و شروع درمان اهمیت دارد.

لغات کلیدی: سندرم نفروتیک مادرزادی، موتاسیون *NPHS2*، آنالیز ژنتیکی

### Article No 18118

#### Mutations in KEOPS-complex genes cause nephrotic syndrome with primary microcephaly Mastane Moghtaderi

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Galloway-Mowat syndrome (GAMOS) is a severe autosomal-recessive disease characterized by the combination of early-onset steroid-resistant nephrotic syndrome (SRNS) and microcephaly with brain anomalies.

Phenotypically, all individuals with mutations in any of the 4 KEOPS genes had primary microcephaly, developmental delay, propensity for seizures, and NS of early onset. Most patients died in early childhood.

Several individuals were noted to have facial dysmorphism sometimes with features of progeria, and skeletal abnormalities such as arachnodactyly.

The KEOPS complex contains 4 subunits LAGE3, OSGEP, TP53RK, and TPRKB. It regulates a universal chemical modification of tRNAs that is necessary for translational accuracy and efficiency.

We thereby discovered recessive mutations in any of the 4 genes encoding KEOPS complex proteins as novel causes of Galloway-Mowat syndrome. We termed these variants of GAMOS due to mutations in LAGE3, OSGEP, TP53RK, or TPRKB, 'GAMOS 2', 'GAMOS 3', 'GAMOS 4' and 'GAMOS 5', respectively. Thus, we made the surprising discovery, that specific recessive mutations in 4 genes that serve a fundamental cellular function cause a distinct renal-neuronal disease phenotype.

### Article No 18119

#### High frequency of metabolite abnormalities in pediatric isolated hematuria

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**Background:** Hematuria, either macroscopic or microscopic, could be originated from anywhere of the kidney and urinary tract and could be a finding of an important underlying disease. Hematuria is always an accidentally finding and the detection of underlying causes are difficult or impossible in most patients. In this study, we evaluated the underlying causes of hematuria in problematic cases.

**Methods:** We included 522 referred children with hematuria who had not renal stone, urinary tract infection or glomerular diseases, based on clinical and laboratory findings. Then we evaluated the records and clinical symptoms and laboratory findings for other structural and hematological disorders and their urinary metabolites in 24-hour urine collection.

**Results:** In this study, from 659 referred children with culture negative hematuria, 100 patients had renal stone in their ultrasonography and 59 patient had findings of glomerular disorders. In other 522 enrolled patient, microscopic and gross hematuria were found in 88.7% and 11.3% respectively, the mean age was 5.9 years and the female to male ratio was 2 to 1. The most common symptoms were occasional abdominal pain and dysuria. Also in the 24-hour urine evaluation, we found metabolic disorders in 94% of cases and the most common findings were hypocitraturia (60.7%) followed by hypomagnesuria (58.2%), hyperuricosuria (35.8%), hypercalciuria (33.7%), hyperoxaluria (24.9%) and cystinuria (in only 4 cases).

**Conclusion:** Disorders in urine metabolites include a large proportion of the underlying factors for hematuria in children and adolescents specially in previously considered cases as idiopathic hematuria. Therefore 24-hour urine collection for urinary metabolites, could be a recommended evaluation in cases with isolated hematuria.

**Key words:** Isolated hematuria, children, urine metabolites

### Article No 18120

#### How many Iranian Cystinosis patients Uses Right Dosage of Cystagon

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**Background:** cystinosis is a metabolic autosomal recessive disorder caused by mutation in cystinosis (CTNS) gene, locus 17p13<sup>1-3</sup> and lead to accumulation of cystine crystals in lysosomes. We conducted guided discussion regarding specific topic, to explore how many of patients use recommended dose of cystagon and how many of them are under treated.

**Methods:** we gathered information on 185 cystinosis patients from the whole country. They were diagnosed by the specialists through eye examination, WBC cysteine level and bone marrow analysis. Among these 185 patients, 40 live in Tehran, 35 in Ahvaz, 18 in Isfahan, 13 in Urmia, 6 in Mashhad, 29 in Shiraz, 10 in Tabriz, 2 in Rasht and 2 in Gorgan.

The recommended dose in these patients was evaluated based on patient's height and body weight. These doses were then compared to the actual daily dose of each patient.

**Results & Discussion:** We found that 42% of patients are treated with lower dose than recommended and among them 26% are taking as low as half of the therapeutic dose. This can be explained by patient's compliance, financial issues and physician's experience. When treatment is optimized (recommended dose for <50 kg is 1.3g/m<sup>2</sup>/day and for >50 kg is 2 g/day), we encounter less side effects and higher life expectancy. Cystagon's dosing should be based on the body surface area and calculated in g/m<sup>2</sup>/day. We strongly suggest our physicians to prescribe Cystagon with the correct dosage for the successful treatment regardless of patients compliance or cost.

### Article No 18121

**Effect of silymarin in reduction of proteinuria in pediatric renal diseases**Shokoo Sharifpoor<sup>1</sup>, Mohammad Ali Momeni<sup>2</sup>

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**Introduction:** Nephrotic syndrome, or nephrosis, is defined as the presence of nephrotic-range proteinuria, edema, hyperlipidemia, and hypoalbuminemia)

*Silybummarianum* is the scientific name for milk thistle (Mt) or St. Mary's thistle is the mixture of mainly three flavonolignans, silybin, silydianin and silychristin

**Methods:** This article is a systematic review through library research and Internet search sites (sid, pubmed, Isc) the 65 articles were collected.

**Results:** Nephrotic syndrome is a consequence of an imbalance between oxidant and antioxidant activity. *Silybummarianum* has properties on renal protection, hypolipidemic and anti-atherosclerosis activities, cardiovascular protection, prevention of insulin resistance that might be effective for prevention of nephropathy-induced premature death in diabetic patients. *Silymarin*-treated group had at least a 50% decrease in urine albumin-creatinine ratio after 3 months of treatment and reduced tumor necrosis factor (TNF)- $\alpha$ , and malondialdehyde in patients with diabetic nephropathy.

*Silybin* has a stimulatory effect on kidney cells by repairing and regenerating increasing via protein and nucleic acid synthesis

**Conclusion:** With respect to multiple side effects of immunosuppressive drugs especially in children who are in process of growing, it's better to consider treatments based on different causative pathophysiologies. Since one of the most important causes of nephrotic syndrome is diabetes mellitus and *silymarin* has hypoglycemic effects and also protects kidney from damages of high blood sugar. The role of oxidative stress in nephrotic syndrome has demonstrated so novel antioxidant therapies including *silymarin* should be considered.

**Key Words:** *silymarin*, proteinuria, antioxidant

**Article No 18122****A difficult case with proteinuria and progressive tremor**

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In this paper, we report a case of interstitial nephritis with proteinuria, neurological manifestations, and uncontrollable blood pressure following chronic thallium poisoning

A 20 years old girl which after a headache, mild hair loss tremor in the upper extremity, high blood pressure and CKD 3 (serum creatinine 1.8mg/dl) and proteinuria +2 in kidney biopsy severe chronic interstitial nephritis with glomerulosclerosis reported) has been hospitalized about 1.5 years ago. In the preliminary studies, she has been diagnosed and treated for Wilson's disease. Although Next study disproved this diagnosis. She has been routinely treated with corticosteroids and antihypertensive drugs (ACE & diuretic & beta blocker). However, due to the lack of favorable response to the increase creatinine to 2mg/dl, and the exacerbation of tremor and hypertension, she has been exterminated for the possibility of a genetic disease which was also disproved. Eventually, study of the patient's case history led us to Heavy metals poisoning which has been confirmed by a urine and blood test for thallium. The special treatment has been started for one month and the patient's contacts were eliminated as much as possible. After a month of treatment, the patient's creatinine was 1.4 and 1+ proteins in urin, while the tremor was incredibly small. Moreover, the patient's blood pressure, which was previously uncontrollable, has been reached 135/85.

- Thallium is a heavy metal odorless and tasteless, it has successfully been used worldwide as a rat poison and ant killer. Because chronic thallium exposure mimics other disease many cases of industrial thallium exposure most likely go unnoticed. Occupational history is important but the patient may not know that he or she has been exposed. On the other hand, accidental poisoning caused by direct contact with and careless handling of thallium-containing materials occurs more frequently
- Thallium is toxic by cumulative intake; it can be absorbed through the skin, respiratory, and GI tracts. Therefore, besides oral ingestion, inhalation of contaminated dust during manufacture, sniffing what was thought to be cocaine, and skin absorption through protective gloves have all been reported as causes of thallium toxicity. In addition, cases of thallium intoxication by intravenous injection of contaminated heroin have been reported. However, the vast majority of cases result from oral exposures.
- In Iran, the most common form of poisoning is exposure to insecticide poisons containing thallium. In Europe and the United States, thallium sulfate was banned from import, export, possession and uses as an agricultural pesticide long ago. Several factors led to its ban, e.g. the invisible traits which makes it easy to be misused, pollutant's effect on non-target toxicity and harming the environment. Given the severe toxicity high chance of human, contamination and its irreparable environmental hazards, we recommend the total removal of this toxic substance.

### Article No 18123

#### Hepatitis B vaccination in pediatric chronic kidney disease and hemodialysis pediatric patients

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Hepatitis B vaccination is recommended for all chronic kidney disease (CKD) pediatric patients. Despite adults, no specific recommendations have been made for higher doses for pediatric hemodialysis patients. However, despite the lack a specific recommendation, some authors believe that the dose of hepatitis B vaccine should be increased for these patients as adults. Anyhow, it should be considered that the response of the immune system in patients with CKD to the vaccines is impaired over time and with aggravation of renal failure .So the patients with progressive kidney disease and patients who are expected to be candidates of the renal transplantation in the future should be vaccinated as soon as possible.

In immunocompetent persons without any risk factor, routine post immunization testing of serum anti-HBs Ab is not necessary ,but serum anti-HBs Ab level should be checked in hemodialysis patients 1-2 months after completion of the primary hepatitis B vaccine series , and if the level is less than 10 mIU / ml, then Hbs Ag should first be checked, and if negative, then the patient should again receive a full series of hepatitis B vaccine, and if the level of anti-HBs Ab is still less than 10 mIU / ml at the end of the second series of the vaccination, the person is considered a non-responder.

Generally, administration of a complete series of hepatitis B vaccine will produce at least two decades of immunity, and it is believed that even if the level of anti-HBs Ab reaches to less than 10 mIU / ml or non-detectable, there is no need to administer a booster dose of vaccine to a healthy person, but serum anti-HBs Ab level of hemodialysis patients should be checked annually, and if it is less than 10 mIU / ml, then administration of a booster dose is indicated.

### Article No 18127

#### PPD before Steroid treatment therapy in nephritic syndrome

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In some reported more than one-third of people in the world are infected and have Latent tuberculosis infection (LTBI) and about 5-10% of them may progress to active tuberculosis (TB) within their lifetime. LTBI is an important diseases and treatment of LTBI in all patients and especially children is very critical. So, screening of all patients that who had the high risk of LTBI such as HIV or in immunodeficient patients are recommended.

Primary nephrotic syndrome (NS) is the most common glomerular disease in children. NS was defined to heavy proteinuria, generalized edema, hyperlipidemia and hypoproteinemia.

All children that have primary NS were treated empirically with high doses of corticosteroids (CS) at presentation and relapses.

Children with NS are immunocompromised patients before starting of CS and be more immune-deficient as a consequence of immunosuppressive therapy because systemic corticosteroid use could decrease systemic immunity. According to impaired defense mechanism of children with NS, the risk of reactivation of LTBI is higher than others and may leading to the occurrence of tuberculosis.

Therefore, it was recommended to done TB screening test in children with NS before starting of CS. There is disagreement about universal or selected cases screening and the method of screening by the tuberculin skin test (TST) or gamma interferon ( $\gamma$ -interferon) for detection of LTBI.

The first method is cheap, easy to perform and was done by every laboratory, but the second method was more expensive and need high technology equipment. Of course, the tuberculin skin test (TST) may have a lower diagnostic value, probably due to this patients are immunocompromised.

### *Article No 18201*

#### **Treatment of CKD-MBD targeted at lowering high serum phosphate and maintaining serum calcium**

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The management of chronic kidney disease-mineral bone disorders (CKD-MBD) strategies develops some major changes during the years, but it seems that the control of serum phosphorus levels is the most important factor to prevent clinical poor outcomes such as MBD and vascular calcification. Progressive decline in kidney function leads to phosphate retention (usually begins in CKD stage 3b) and decrease in the production of active vitamin D and increase parathyroid hormone (PTH) and fibroblast growth factor - 23. PTH and FGF-23 have same effect in decreasing phosphorus level. A key question rises here is which level of serum phosphorus in CKD stages can prevent these clinical outcomes?

Dietary phosphorus restriction, phosphate binder therapy and active vitamin D supplementation and adequate scheme of dialysis are the most important strategies for this goal of therapy.

Today, although most centers decide to control serum phosphorus when the hypophosphatemia begin to happen, there are some evidences that it might be better to start to control serum phosphorus before the onset of hyperphosphatemia. Earlier phosphorus control and achieving normal phosphorus levels may help to reduce clinical consequences of CKD-MBD. But unfortunately control of serum phosphorus level is very difficult and hyperphosphatemia continues to be extremely common in CKD patients.

**Key words:** chronic kidney disease, MBD, hyperphosphatemia, phosphate binders

### *Article No 18203*

## Prevalence of hepatitis B and C infection among hemodialysis children and adolescents in Iran: a single center experience

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**Background and Aims:** It is well known that patients undergoing dialysis treatment and in particular hemodialysis (HD), are at increased risk of acquiring hepatitis B virus (HBV) and hepatitis C virus (HCV) infection due to high number of blood transfusion sessions, prolonged vascular access, high exposure to infected patients, and contaminated HD equipment during treatment. Few data are available on the prevalence of HBV and HCV infection among HD children. The aim of this study was to determine the prevalence of hepatitis B and C infection in children with end stage renal disease (ESRD) on HD.

**Materials and methods:** In this cross-sectional study, all children and adolescents with ESRD ( $n = 149$ ) dialyzed at Children's Hospital Medical Center between January 1991 and December 2009 were enrolled into the study. The patients' demographic data, clinical characteristics, and treatment modalities were extracted from hospital profiles of admitted patients. All children who dialyzed in the center were routinely screened for the presence of **hepatitis B** surface antigen (HBsAg), anti-HCV antibody at the initiation of dialysis treatment. HBsAg and HCV antibodies were measured using specific enzyme-linked immunoassay kits as serological testing. All patients with HIV infection were excluded from the study.

**Results:** A total of 149 children (51% male and 49% female) with ESRD were referred for HD treatment during the study period. The mean age of children was 8.80 years (range 3 months to 18 years). Two (2.04%) patients were HBsAg positive, 2(2.04%) were anti HCV positive. **Co-infection** with both HBV and HCV was found in three (2.72%) HD children. The prevalence of HBV was equal to HCV.

All patients with positive HBV and HCV or with **HBV/HCV co-infection** were clinically asymptomatic. Glomerulopathies (22.8%) and reflux nephropathy (16.1%) were the most common causes of end stage renal disease in studied patients. Eighty-two children (57.04%) **underwent HD** three sessions per week, **32 (21.47%)** two sessions per week, **33 (22.14%)**, one session per week and two (1.34%) five sessions per week. The mean duration of HD was 34 months. Death occurred in 18 patients (12%), mainly due to cardiovascular and infectious complications.

**Conclusions:** Prevalence of HBV and, HCV in hemodialysis children seems low. However, HBV DNA (occult hepatitis B virus infection) was not evaluated in children with negative test for HBsAg, which needs further investigation. HBV vaccination, implementations of strict infection-control program are highly recommended for preventing the transmission of infection in the hemodialysis units.

**Keywords:** Prevalence, HBV, HCV, Hemodialysis, Children, End Stage Renal Disease

### Article No 18204

## Treatment of abnormal PTH levels in CKD-MBD

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### STAGES OF CHRONIC KIDNEY DISEASE:

- Stage 1 – Normal glomerular filtration rate (GFR)  $\geq 90$  mL/min per 1.73 m<sup>2</sup>
- Stage 2 – GFR between 60 to 89 mL/min per 1.73 m<sup>2</sup>
- Stage 3 – GFR between 30 and 59 mL/min per 1.73 m<sup>2</sup>

- Stage 4 – GFR between 15 and 29 mL/min per 1.73 m<sup>2</sup>
- Stage 5 – GFR of <15 mL/min per 1.73 m<sup>2</sup> or requires dialysis treatment

Children with CKD stage 2 usually have no signs or symptoms of bone abnormalities. However, these children may have evidence of abnormalities on laboratory testing (eg, decreased serum calcitriol and elevated serum parathyroid hormone [PTH]); this period should be used to educate the child and family about CKD and its impact on bone metabolism.

Subtle signs of renal osteodystrophy begin to be observed when the GFR decreases to 50 percent of normal (stage 3 diseases), these children should be monitored for evidence of bone disease by physical examination and laboratory evaluation, and Physical findings include muscle pain, weakness, and bony changes such as varus and valgus deformities of the long bones.

Laboratory abnormalities of bone metabolism (eg, elevated PTH) are common in stage 3 disease and require therapeutic interventions.

**KDOQI** guidelines in the management of bone metabolic abnormalities in children with CKD include the following:

- Management (frequency of monitoring and therapeutic interventions) is based upon the child's level of kidney function.
- Therapy focuses on the prevention of phosphate retention and hypovitaminosis D (development of secondary hyperparathyroidism, which results in renal osteodystrophy).
- Suggest that serum concentrations of calcium, phosphate, and parathyroid hormone should be measured on an ongoing basis in all children with stages 2 to 5 of CKD
- In children with an elevated serum PTH and/or phosphate level, we recommend reducing the PTH and/or phosphate concentration with dietary restriction of phosphorus and the use of phosphate binders, if necessary.
- In children with CKD stage 2 to 4, if serum PTH is above the target range, 25-hydroxyvitamin D concentration should be measured. If the 25-hydroxyvitamin D level is <30 ng/ml, we suggest that ergocalciferol or cholecalciferol be given, If the level is >30 ng/ml and the serum calcium level is <10 mg/dL, we recommend calcitriol therapy.
- In children with CKD stage 5 and PTH levels >300 pg/mL, we suggest that calcitriol should be administered until serum PTH is reduced to a range between 200 to 300 pg/mL, although the optimal target range remains controversial.
- Serum calcium should be maintained within a normal range for the laboratory used, generally between 8.8 and 9.7 mg/dL, if the patient is hypocalcemic, we recommend that calcium supplementation and/or vitamin D therapy be given.

**MANAGEMENT:** The goals of therapy are to prevent phosphate retention, hypovitaminosis D, and hypocalcemia. Management is focused on early detection and correction of these abnormalities because each contributes to the development of secondary hyperparathyroidism, which causes renal osteodystrophy. In children with CKD, two therapeutic interventions include restriction of dietary phosphorus and the use of calcium-based phosphate binders. Dietary phosphorus should be restricted to 80 percent DRI if the serum PTH is above the target range and the serum phosphate concentration is above the age-appropriate normal range. Phosphate-binding compounds will be effective in lowering serum phosphate levels only if dietary phosphate restriction is continued. Phosphate binders should be taken 10 to 15 minutes before or during the meal.

Phosphate binders, regardless of the agent used, have a limited phosphate-binding capacity. As examples, 1 g of **calcium carbonate** binds 39 mg of phosphate, 1 g of **calcium acetate** binds 45 mg of phosphate, and 400 mg of **sevelamer HCl** binds 32 mg of phosphate.

Noncalcium and nonaluminum phosphate binders, such as **sevelamer** and **lanthanum carbonate**, are used more frequently as initial therapy in adults.

Sevelamer HCL was associated with a reduction in LDL cholesterol and the frequency of hypercalcemic episodes, but an increase in the frequency of metabolic acidosis.

Sevelamer carbonate (Renvela) was subsequently found to be as effective as a phosphate binder compared to sevelamer HCL.

Lanthanum carbonate is a more potent binder of intestinal phosphate than other current binders. However, tissue accumulation of lanthanum has been shown in rats and increased bone lanthanum levels have been seen in adult subjects .

The following phosphate binders should be **avoided** in children with CKD:

- Aluminum hydroxide.
- Calcium citrate.
- Magnesium-containing antacids

**Potential role for FGF23:** Serum FGF23 is emerging as a potential treatment target for management of CKD-MBD.

**Vitamin D:** vitamin D therapy improves bone disease as measured by parathyroid hormone levels, regardless of formulation or route of administration.

The KDOQI guidelines recommend that treatment with ergocalciferol or cholecalciferol should be started when the serum 25-hydroxyvitamin D is <30 ng/mL.

In children with CKD stage 2 to 4, active vitamin D analogue (eg, calcitriol) should be considered if all of the following criteria are met:

- Serum 25-hydroxyvitamin D (25[OH]D) is >30 ng/mL.
- Serum PTH is above the target range
- Serum calcium level is <10 mg/dL.
- Serum phosphorus level is less than the age appropriate upper limits for the stage of CKD

If secondary hyperparathyroidism persists despite normal serum phosphorus and vitamin D levels, treatment with a vitamin D sterol or vitamin D analog is indicated. Calcitriol and its prohormone, alfacalcidol, are widely used in children, and both are effective in suppressing PTH by increasing intestinal calcium absorption and suppressing PTH gene transcription.

**Calcimimetics** treat hyperparathyroidism by binding to the calcium-sensing receptor of the parathyroid gland and, through allosteric modification of the receptor, increase its sensitivity to ionized calcium.

**Cinacalcet**, the only currently available calcimimetics agent, has been shown to effectively lower serum PTH levels in adult ESRD and CKD patients

**CKD stage 5:** In children with CKD stage 5 disease and serum PTH >300 pg/mL, calcitriol should be administered to reduce the serum PTH to 200 to 300 pg/mL according to the KDOQI guidelines.

Once calcitriol is started, serum calcium and phosphorus concentrations should be measured every two weeks for one month and at least monthly thereafter.

Serum PTH should be measured monthly for three months and then at least every three months. Serum PTH concentrations <100 ng/L should be avoided to prevent adynamic bone disease. Based KDOQI guidelines recommend targeted levels of serum intact PTH at different stages of CKD as follows:

- Stage 2 and 3 disease – 35 to 70 pg/mL
- Stage 4 disease – 70 to 110 pg/mL

Stage 5 disease – 200 to 300 pg/mL

Intake of calcium for children with CKD is twice the daily dietary recommended intake for age, which may easily be exceeded with large calcium-based binder dosages.

**Hypocalcemia:** The total serum calcium should be maintained within a normal range for the laboratory used, generally between 8.8 and 9.7 mg/dL.

**Hypercalcemia:** If the total serum calcium value exceeds 10.2 mg/dL, the dose of calcium-based phosphate binders should be reduced and/or therapy changed to **sevelamer**. Vitamin D therapy should also be discontinued until the serum calcium returns to the targeted range and then restarted with an appropriate dose adjustment.

**Surgical care:** When refractory hyperparathyroidism develops despite aggressive pharmacotherapy (persistent serum levels of intact parathyroid hormone greater than 800 pg/mL, associated with hypercalcemia and/or hyperphosphatemia that is refractory to medical therapy), total parathyroidectomy

with autotransplantation to the forearm or abdomen may be considered a safe and effective alternative in pediatric patients. This approach to management is usually considered a last option because of the resultant potential difficulties with calcium homeostasis following renal transplantation.

Indications for surgery include bone pain or fracture, pruritus, calciphylaxis, and extraskelatal nonvascular calcifications with elevated parathyroid hormone levels despite appropriate medical therapy.

### *Article No 18205*

## **Hypoparathyroidism versus hyperparathyroidism in pediatric dialysis patients; a single center study**

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**Introduction:** Abnormalities in calcium, phosphorous and parathyroid hormone (PTH) metabolisms are common in dialysis patients. Reaching target levels for these serologic factors and calcium × phosphorous products is recommended to minimize cardiovascular events.

**Objectives:** The aim of this study was to examine calcium, phosphorous and intact PTH (iPTH) abnormalities in a group of dialysis patients.

**Patients and Methods:** Bone minerals status and iPTH levels were assessed in 46 dialysis patients aged 19-300 ( $165.2 \pm 75.73$ ) months. Low and high Ca dialysate solutions routinely were used for hemodialysis (63%) and peritoneal dialysis (30.4%) patients respectively. Comparisons between groups were performed with considering age ( $\leq 5$ , 6-10, and  $> 10$  years), gender and modality of dialysis.

**Results:** Serum calcium and corrected calcium levels were significantly higher in peritoneal dialysis (PD) patients. Hypoparathyroidism was the most frequent iPTH abnormality (58.7%). It was more prevalent in males. Hyperparathyroidism was more frequent in females.

**Conclusions:** We found that hypoparathyroidism is the most prevalent PTH abnormality. We also noted that patients on peritoneal dialysis are more prone to develop this form of PTH abnormality. We found that phosphate control is better in peritoneal dialysis vs. hemodialysis cases.

**Keywords:** CKD, Hemodialysis, Peritoneal dialysis, PTH

### *Article No 18208*

## **Vascular access challenges in children with CRF**

Banafshe Dormanesh

The incidence and prevalence of ESRD in pediatrics had increased. As we know, like adults Hemodialysis is the major first line of renal replacement therapy . So for almost all pediatrics with stage 4 CKD we need a vascular access. The ideal vascular access delivers an adequate flow rate in hemodialysis procedure, also has a long use-life with the lowest incidence of complications .

We usually select one of these 3 types of vascular access: arteriovenous fistula (AVF) , arteriovenous graft (AVG) , central venous catheter (CVC)

Selection the method depends on many factors such as patients age and their physical condition and distance to the start of the first dialysis session , parental cooperation and acceptance .

In this article, we will compare different methods and express the advantages and disadvantages of each of them while pointing briefly to the prerequisite and efficiency of each method. We also express operational strategies (including all personal, family and nursing medical care's) to increase their efficiency, reduce and delay complications .

### *Article No 18209*

### Effect of allopurinol in slowing kidney disease progression in hyperuricemic children with CKD stages 1-3

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**Background** Hyperuricemia is a leading risk factor for the development of chronic kidney disease (CKD). We hypothesized that lowering serum uric acid (SUA) with allopurinol in hyperuricemic children with CKD may reduce the risk of CKD progression.

**Patients and Methods** A total of 91 children, aged 3-15 years, with elevated serum uric acid level (SUA) >5.5 mg/dL and CKD stages 1-3 were prospectively randomized to receive allopurinol 5mg/kg/day (study group, n=49) or no treatment (control group, n=42) for 4 months. The primary and secondary outcomes were changes in estimated glomerular filtration rate (eGFR) (>10 mL/min/1.73m<sup>2</sup>) and the SUA (>1.0 mg/dL) from baseline values, respectively.

**Results** Baseline age, gender, blood pressure (BP), body mass index (BMI), SUA, high-sensitive C-reactive protein (hsCRP), and eGFR were similar in allopurinol subjects and controls. Allopurinol treatment resulted in a decrease in SUA, a decrease in systolic and diastolic BP, a decrease in hsCRP and an increase in eGFR compared with the baseline values (p<0.05 for all). No significant difference was observed in the control hyperuricemic subjects. In multiple regression analysis after incorporating variables (age, gender, BMI, systolic and diastolic BP, CRP, and SUA), eGFR was independently related to SUA both before and after treatments (p=0.03 vs. p=0.02, respectively). All patients in the study group tolerated allopurinol, and there was no adverse reactions observed by physical examination or reported by patients.

**Conclusion** Treatment of hyperuricemia with Allopurinol in children with CKD stages 1-3, over a 4 month-period was associated with increased eGFR, lower SUA levels, and hsCRP.

**Keywords:** Allopurinol, Children, Chronic Kidney Disease, High-sensitive C-reactive protein, Hyperuricemia

### Article No 18211

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### BK nephropathy in pediatric renal transplantation

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BKV associated nephropathy is an important and common complication after renal transplantation. BKV nephropathy is a significant risk factor of graft dysfunction and an important cause of graft loss. The risk factors of BKV associated nephropathy are younger age of recipients, HLA mismatching, the number of acute rejection episodes, the presence of delayed graft failure, increased cold ischemia time, induction by tymoglobuline and use of mycophenolate mofetil with tacrolimus in maintenance therapy. Over the past years, the studies focused on the preventive protocols. Ciprofloxacin has not any effect to prevent BKV use of brincidofovir, 'lipid oral formulation of cidofovir', is effective as prophylaxis. BKV vaccine is also under research. Treatment options are limited and controversial. The mainstay treatment is immunosuppressive reduction. Discontinuation of mycophenolate mofetil is a common approach. Conversion of tacrolimus to cyclosporine is another approach. There are other therapeutic options such as the use of leflunomide, cidofovir, IVIg and everolimus.

### Article No 18212

### **Efficacy and safety of rituximab in children with difficult-to-treat nephrotic syndrome; a systematic review**

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**Objectives:** Different types of studies have been done so far on drugs efficacy and safety in children with refractory nephrotic syndrome. Ofatumumab might be an effective treatment for this syndrome; however, the long-term effects and cost-effectiveness of Ofatumumab treatment have not been comprehensively assessed. This study aims to do a systematic review on the efficacy and safety of Ofatumumab in children with difficult-to-treat nephrotic syndrome.

**Material and Methods:** An electronic literature search was conducted to identify appropriate studies. The search term was: (“nephrotic syndrome” or “minimal change disease” or “focal segmental glomerulosclerosis” or membranous) and (“Ofatumumab” or “CD20” or “Arzerra” or “HuMax-CD20”). We included all studies related to using Ofatumumab in children with difficult-to-treat nephrotic syndrome. Two independent reviewers extracted data from the articles according to the selection criteria.

**Results:** Eligible studies were included in this systematic review. The literature search and reference mining yielded 77 potential relevant articles. We removed 32 articles because of duplication. Also, 26 references were excluded after reviewing the titles and abstracts because they were books, book sections, review papers and therefore not relevant. Then, we reviewed full-text of selected articles and removed 14 other studies because the topics were not relevant to the subject. At last, 5 studies were included in the systematic review. The metric considered to assess the efficacy of Ofatumumab in children with nephrotic syndrome in most of the studies was complete remission rate.

**Conclusions:** Our systematic review shows that Ofatumumab may be effective in the treatment of refractory nephrotic syndrome in children, and it can reduce the use of steroid and immunosuppressants. However, acknowledging the limitations of the study due to the size and nature of the studies included, further large randomized trials are suggested.

**Keywords:** Ofatumumab, Nephrotic syndrome, Children, Treatment

#### **Article No 18214**

### **Incidence of malignancy after living kidney transplantation: a multicenter study from iran**

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Malignancy is a common complication after renal transplantation. However, limited data are available on post-transplant malignancy in living kidney transplantation. Therefore, we made a plan to evaluate the incidence and types of malignancies, association with the main risk factors and patient survival in a large population of living kidney transplantation. We conducted a large retrospective multicenter study on 12525

renal recipients, accounting for up to 59% of all kidney transplantation in Iran during 22 years follow up period. All information was collected from observation of individual notes or computerized records for transplant patients. Two hundred and sixty-six biopsy-proven malignancies were collected from 16 Transplant Centers in Iran; 26 different type of malignancy categorized in 5 groups were detected. The mean age of patients was  $46.2 \pm 12.9$  years, mean age at tumor diagnosis was  $50.8 \pm 13.2$  years and average time between transplantation and detection of malignancy was  $50.0 \pm 48.4$  months. Overall tumor incidence

in recipients was 2%. Kaposi's sarcoma was the most common type of tumor. The overall mean survival time was 117.1 months (95% CI: 104.9-129.3). In multivariate analysis, the only independent risk factor associated with mortality was type of malignancy. This study revealed the lowest malignancy incidence in living unrelated kidney transplantation.

Key words: malignancy, kidney transplantation, incidence.

#### *Article No 18215*

### **BK virus-associated hemorrhagic cystitis in children suffering malignancies: the report of three cases**

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BK virus-associated hemorrhagic cystitis is frequent in patients who undergoing bone marrow transplantation but it rarely occur among other immunosuppressed patients particularly in those who are planned for chemotherapy due to hematopoietic malignancies. Herein, we describes our experience on three children with burkitt lymphoma, leukemia and astocytoma undertreated with chemotherapy suffering HC due toBK virus infection that was successfully controlled and resolved. It can be concluded that by early detection of BK virus using RT-PCR technique and then administration of antiviral agents especially IVIG or leflunomide, BK virus-related HC can be successfully managed in children affected by malignancies that their treatment by chemotherapy can be even continued after virus elimination.

#### *Article No 18217*

### **Encapsulate Peritoneal Dialysis in Peritoneal Dialysis -short term post PD**

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Encapsulated Peritoneal Sclerosis (EPS) is a devastating complication of long term CAPD. The diagnosis is based on structural and functional aspects of intestinal obstruction. The total imaging score at the time of diagnosis of EPS did not correlate with the clinical outcome. It is important to differentiate simple

peritoneal sclerosis from EPS. The incidence increases from zero to 18% with time on peritoneal dialysis for 5-8 years. The risk of EPS increases exponentially when PD continues beyond 3 years. The other potential risk factors are high strength glucose exposure, icodextrin, young age, inflammation, chemical exposure, genetic factors, acidic PD fluid. Peritoneal injury and subsequent peritoneal inflammation are two hit hypothesis for EPS. But episodes of peritonitis, intense or repeated hemoperitoneum, abdominal surgery, stopped PD, and genetic predisposition could be the potential risk factors. There is no authentic screening tool for early diagnosis. The combination of Ca-125 < 33 U/min and IL-6 > 350 pg/min with UFF suggest the possibility to identify patients at risk. High levels of cytokines in peritoneal effluent correlate with alteration peritoneal membrane transport status. The pathophysiology of EPS consists of inflammation, fibrin deposition and fibrinolysis, epithelial-mesenchymal transition, and growth factors. Ultrafiltration failure and high average transport status are very common in EPS. High awareness to detect the earliest stage of EPS might help to improve survival. Discontinuation of PD, nutritional support, immunosuppressive therapy, tamoxifen and surgery are medical options. There is no strategy to prevent EPS. In the case of PD catheter removal, dry peritoneum might lead to new fibrin deposition and accelerate sclerosing process. Periodic irrigation of peritoneal cavity for 6-12 months after cessation of PD therapy might prevent intestinal adhesion.

### The efficacy of Icodextrin in children on Peritoneal Dialysis (Ali Asghar Hospital Report )

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**Introduction:** Peritoneal dialysis (PD) is a therapy that typically is managed by patients at home. The therapy works by cleaning the blood of toxins and removing extra fluids through one of the body's own membranes, the peritoneal membrane. Extraneal (icodextrin 7.5%) is recommended as a once daily replacement for a single glucose exchange as part of a continuous ambulatory peritoneal dialysis (CAPD) or automated peritoneal dialysis (APD) regimen for the treatment of chronic renal failure, particularly for patients who have lost ultrafiltration on glucose solutions, because it can extend time on CAPD therapy in such patients.

**Method :** in this retrospective cohort study in ali asghar hospital in nephrology unit we studied 22 patients under peritoneal dialysis with icodextrin solution in 2014

**Results:** From 22 patients including 13 female & 9 male that 18 patient of them was chronic and 4 was acute diseases. the beneficial effect of icodextrin compatible with other solutions is :

- Increase ultrafiltration
- decreased Edema
- Increasing patients satisfaction with the treatment process due to dwell time
- Improving of control Blood pressure

**Conclusion:** This study further supports previous findings of long-term utilization of icodextrin solution improving patient and technique survival in PD patients.

### Article No 18218

### Urinary Calprotectin as a Marker to Distinguish Functional and Structural Acute Kidney Injury in Pediatric Population

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**Introduction:** Acute kidney injury (AKI) is a serious, common and occasionally under-recognized condition. To date, the clinical and some laboratory parameters are routinely applied to distinguish between functional and structural AKI which can be challenging in certain occasions. In the present paper, we investigate the accuracy of urinary calprotectin as a diagnostic biomarker in this dubious situation.

**Methods:** This is a cross-sectional study among 75 children with AKI defined by Acute Kidney Injury Network (AKIN) and 20 healthy children as controls which was carried out for about six months (September 2014 to March 2015). Random urinary calprotectin concentration was assessed by ELISA in both groups within 48 hours after diagnosis. Patients with obstructive uropathy, malnutrition, renal transplantation, chronic renal failure, urinary tract infection, and malignancy were excluded. Receiver-operating characteristic (ROC) curves were drawn to determine the accuracy of urinary calprotectin to detect children with structural AKI. P value less than 0.05 was considered significant.

**Results:** Median urinary calprotectin was 1240 ng/mL in structural AKI, 28.5 in functional, and 33 in controls. Receiver operating curve analysis revealed high levels of accuracy for measuring calprotectin in predicting structural AKI. A cutoff level of 230 ng/mL for urinary calprotectin showed high sensitivity and specificity. The urine calprotectin/creatinine ratio indicated the same accuracy as urinary calprotectin in diagnosing structural AKI. The ROC curve function was better for urine calprotectin and its ratio in comparison to fractional excretion of sodium (FENa).

**Conclusions:** Calprotectin is a biomarker that can rapidly and easily recognize structural from functional AKI with high sensitivity and specificity in comparison to traditional most accurate diagnostic test; FENa.

### Article No 18220

#### The association between matrix metalloproteinases-2, 9 and endothelial dysfunction with echocardiographic findings in children with end stage renal disease

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**Introduction:** Cardiovascular disease (CVD) is the most likely cause of mortality in children with end stage renal disease (ESRD). Matrix metalloproteinases-2, 9 (MMP-2, 9), and endothelial dysfunction could contribute in CVD development in such patients. The aim of this study was to evaluate the association between MMP-2, 9 and endothelial dysfunction markers (sE-selectin and brachial flow mediated dilatation (FMD)) with echocardiographic findings in children with ESRD.

**Methods:** 31 children with ESRD and 18 healthy age- and sex- matched subjects as controls were recruited. Serum levels of MMP-2,9 and sE-selectin were measured. Brachial FMD and echocardiographic parameters were evaluated.

**Results:** Serum levels of MMP-2, 9, and sE-selectin were significantly higher in patients than controls. The means of brachial FMD, ejection fraction (EF), and E/A ratio were significantly lower in patients than controls. Children with ESRD had higher means of left ventricular end systolic diameter (LVESD) and left ventricular end diastolic diameter (LVEDD) than controls. MMP-2 was correlated with EF, and E/A ratio negatively and was positively correlated with LVES, and LVED. MMP-9 was inversely correlated with EF.

**Conclusion:** MMP-2, 9 were associated with cardiac abnormalities in children with ESRD. We didn't find correlation between endothelial dysfunction markers and echocardiographic parameters.

**Key words:** Matrix metalloproteinase-2 (MMP-2); Matrix metalloproteinase-9 (MMP-9); sE-selectin; Flow mediated dilatation (FMD); End stage renal disease (ESRD); Cardiovascular disease (CVD); Children; Echocardiography.

### Article No 18221

### Satiety Hormones in Pediatric Urinary Tract Infection

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**Introduction:** Urinary tract infections (UTIs) are among common infections in pediatric patients and are associated with anorexia and failure to thrive (FTT) although transient. Appetite regulating hormones may be considered as a causative factor. Leptin is a 16kd protein, secreted by adipocytes, regulates appetite, food intake and body weight via hypothalamic melanocortin-4 receptor (MC-4R). Leptin is also a potential cytokine for inflammation.

Ghrelin on the other hand is a regulatory hormone that maintains fat tissues and body composition. Ghrelin is mainly produced in stomach but is also produced in smaller amounts in other organs such as kidneys.

Ghrelin stimulates release of growth hormone (GH), increases food intake, and causes weight gain, mainly in fat tissue. Ghrelin increases body fat without extra calorie intake. It is postulated that Ghrelin may make metabolism more efficient.

Ghrelin inhibits Leptin receptor, an internal ligand for the growth hormone secretion (GHS-R), so it is likely that there is an association between infection and Ghrelin.

The aim of this study was to evaluate serum and urine Leptin and Ghrelin before and after treatment in children with UTI.

**Material and Methods:** A cross-sectional study was performed on 104 children between 2 months to 17 years old who were admitted in nephrology ward of Mofid hospital. They were evaluated by history, physical examination, blood tests and urine analysis, ultrasound, nuclear scan, and in some cases kidney biopsy according to the Glomerular Filtration Rate (GFR) and KDOQI guidelines and instructions. Patients were divided into 5 groups based on CKD stages: results were expressed using descriptive statistics differences considered statistically significant if  $p < 0.05$ .

**Results:** Of the 104 cases, 56 patients (45%) were male and 48 (46%) were female. The age range was  $86.6 \pm 9.4$  months.

High systolic or diastolic blood pressure was present in 37.5% of patients, anemia was seen in 74%, 70% had bone disease and 63% had failure to thrive. The most common etiology of chronic kidney disease was neurogenic bladder and reflux nephropathy in 27.8% and 16.34%, respectively.

Twenty percent of patients were in stage 1 CKD, 24% at stage 2, 17% at stage 3, 40% were at stage 4 and 5 and 31.7% of patients had a GFR less than 15 at the time of diagnosis and 11.53% of patients treated with renal replacement therapy (RRT). During the study, 4.8% of patients received a kidney transplant.

**Conclusion:** As renal complications such as hypertension and end stage renal disease are high in neurogenic bladder, this entity should be considered in etiology of CKD in children. Early recognition and management of CKD complications is of utmost importance for improvement of quality of life of these children.

**Key words:** Renal insufficiency, Complication, Hypertension, Anemia, Bone disease, Failure to thrive, Children.

### Article No 18222

### Correlation of severity of prenatal hydronephrosis and severity of vesicoureteral reflux (with a review on Hydronephrosis Algorithmic approach)

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**Introduction:**Hydronephrosis is one of common congenital abnormality detected prenatally by ultrasonography .there is a lot s of controversy about doing post natal cystography for these patients. this study was done for determine correlation of degrees of prenatal hydronephrosis and degrees of vesicoureteral reflux(VUR).

**Method:** Infants, whose antenatal ultrasonography (US) showed a fetal renal pelvic diameter(RPD) of 5 mm or greater were investigated. Prenatally hydronephrosis severity was defined base on 3rd trimester sonography according to RPD defined as mild(5-9mm), moderate(10-14mm) or severe(>15mm).VUR degree were classified as mild (grade &2),moderate(grade 3),severe(grade 4&5).

Postnatal urinary tract sonography and cystography were performed for all of them and according proposal algorithm, DMSA scan, DTPA scan were done if were indicated.

**Result:**A total of 200 infants were enrolled in this study. Each kidney detected to have reflux was referred to as a “renal unit”(RU) and a total of 400 RUs were investigated. There were 143 (71.5%) males and 57(28.5%) females . mean of APD of prenatal hydronephrosis were 8 milimeter +/-6 (maximum =60 mm).There was a significant but weak correlation coefficient between degree of prenatal hydronephrosis and degree of VUR(p.value <0.001; R=0.22; spearman test).also , the same result was shown between degree of postnatal hydronephrosis and degree of VUR(p.value <0.001; R=0.3; spearman test).

**Conclusion:** VUR is a relatively common finding in neonates referred for hydronephrosis. We recommend cystography for all of neonate with moderate and severe prenatal hydronephrosis and for neonate with mild prenatal hydronephrosis with other reported abnormality in kidney or bladder in sonography,increase RPD in postnatal sonography,history of urinary tract infection or sibling VUR or history of abnormal voiding.

### Article No 18223

#### Is There a Correlation Between Anteroposterior Renal Pelvic Diameter (APD) and Vesicoureteral Reflux

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**Introduction:** One of the most common reasons for kidney evaluation in neonates and infants is congenital hydronephrosis. The correct diagnosis (i.e. VUR or obstructive uropathy or transient hydronephrosis) and its treatment is of great value. This study was conducted to investigate the correlation between APD and VUR in congenital hydronephrosis.

**Material and method:**All neonates with congenital hydronephrosis were investigated by ultrasonography. Neonates with hydronephrosis persisted after birth underwent voiding cystourethrography (VCUG). If VUR was detected, the next step was DMSA scan; otherwise DTPA scan was done to rule out obstructive uropathy. All cases were followed for at least 6 months to confirm the final diagnosis.

**Results:** Out of 89 cases, 73 (82%) were male. There were 8 (8.98%), 36 (40.44%), 45(50.56%) cases of bilateral, right side and left side hydronephrosis, respectively. There were 20 (22.47%) cases of vesicoureteral reflux. Other diagnosis included ureteropelvic junction obstruction (UPJO), ureterovesical junction obstruction (uvjo) and transient hydronephrosis in 23(25.84%), 3(3.37%) and 43(48.31%) of cases, respectively.

Mean APD was  $17.85 \pm 11.71$  totally, with minimum 4mm and maximum 60 mm. Mean APD in patients with VUR was  $12.15 \pm 5.68$  and  $19.95 \pm 13.06$  in cases without VUR. Mann-Whitney U test showed a significant correlation between APD and VUR, outcome (surgery or resolving) and APD with P value of 0.008 and 0.0001, respectively. In addition, Kruskal-Wallis test showed significant correlation between APD and final diagnosis (P value= 0.002).

**Conclusion:** Postnatal APD in ultrasonography has significant correlation with the final diagnosis (including VUR, UPJO, UVJO and transient), outcome and VUR. APD could be valuable in the diagnosis of congenital hydronephrosis.

**Keywords:** congenital hydronephrosis, neonates, ultrasonography, vesicoureteral reflux

### Article No 18224

#### Interaction between Gentamicin and Mycophenolate Mofetil In Experimentally Induced Pyelonephritis

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Acute pyelonephritis (APN) is an inflammatory disease that leads to kidney malfunction. The objective of this investigation was to evaluate the impact of gentamicin (GEN) and ceftriaxone (CEF) alone and in combination with mycophenolatemofetil (MMF) on experimentally induced APN. Forty two Wistar male rats were assigned into seven groups +APN, APN +GEN, APN+CEF, APN+MMF, APN+GEN+MMF and APN+CEF+MMF. APN was induced by injecting *E. coli* in the left kidney. The control and +APN groups were treated with normal saline while the other APN groups received GEN, CEF, or MMF alone and/or in combination for 2 weeks. The elevated total white blood cells count and increased level of creatinine and blood urea nitrogen (BUN) in +APN groups returned to normal levels following 14 days treatment with GEN and CEF. Co-administration of GEN with MMF could not recover the APN-induced changes and resulted in a significant ( $P < 0.05$ ) elevation of creatinine and BUN levels. Histopathological studies supported the biochemical findings as GEN and CEF alone could partly restore the APN-induced degeneration and leukocytic infiltration; however, the combination therapy of GEN plus MMF failed to reduce the APN-induced damages. The antibacterial susceptibility test demonstrated that the strain of *E. coli* used in this study was susceptible to GEN and CEF and the combination therapy did not change the antibacterial potency. These findings suggest that co-administration of GEN with MMF in APN may enhance kidney damage and the adverse effects of combination therapeutic regimen could be related partly to incompatibility of these compounds.

**Keywords:** Acute pyelonephritis, ceftriaxone, combination therapy, *E. coli*, gentamicin, mycophenolatemofetil

### Article No 18226

#### Renal ultrasonography set up in children

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Sonography of the kidneys is very important in the diagnosis and management of renal diseases. The kidneys are examined without any radiation exposure and many pathological changes in the kidneys can be diagnosed with ultrasound, in addition real time sonography is a valuable measure for successful renal biopsy. There are many different types of ultrasound machines but basic principles are the same. For performing renal sonography a clinician should be familiar with types of sonography probes and their application, each knob and control on machine such as gain control, depth, focus, zoom, freeze, save, cine (clip) to get the best possible image. In addition a clinician must know different scan planes (longitudinal, transverse), proper probe handling, positioning the patient and normal appearance and non pathologic findings of renal sonography. In this panel we are going to show practically how a bedside renal sonography is set up and done for those who have not had any previous experience with performing sonography in order to take better patient care in both diagnosis and interventional aspects.

### Article No 18227

#### How to Perform Kidney Biopsy under Ultrasound Guidance

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Renal biopsy is an invaluable diagnostic method in nephrology subspecialty field. It has diagnostic and prognostic values. As a diagnostic method it was done a century ago by indirect visualization of kidney. The method of kidney tissue sampling has been developing in two ways: 1. From blind sampling to more developing type which has been taken under real-time direct visualizing by ultrasound guidance 2. Use of manual to Co-Axial automatic biopsy needles(guns).

To minimized major complications such as hemorrhage and hematoma, a history, physical examination, appropriate laboratory tests should be performed and make a point for exact needle insertion and any kidneys abnormalities by pre-biopsy kidneys ultrasound.

As a general rule patient should be fix in prone position and put an appropriate towel under biopsy side. Then marked area should prep and drape, use local anesthesia.

There are two maneuvers for kidney biopsy of lower pole of kidneys (Usually left kidney): Short-Axis (Out-of-Plane) versus Long-Axis (In-Plane) visualization. If available the coaxial needle can be easily positioned at the site of kidney biopsy for extracting more tissue with less damage.

This brief lecture will illustrate Short-Axis (Out-of-Plane) versus Long-Axis (In-Plane) visualization and taking samples by Co-axial automatic gun on a special phantom model.

### Article No 18229

#### Catheter insertion with ultrasonography guide

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Central venous catheter (CVC) placement is frequently required in the care of critically ill and injured children. Common indications for placement include reliable venous access for medication administration, monitoring of central venous pressure and central venous oxygen saturation, parenteral nutrition, and frequent blood sampling. CVCs are also placed for hemodialysis, hemofiltration, and apheresis in the PICU.

Three sites are commonly used for paediatric CVC placement: femoral, internal jugular, and subclavian. Most CVCs are placed using the wire-guided (Seldinger) technique, in which a needle or catheter-over-needle unit is introduced into the desired vein, blood is aspirated, and a guide wire is placed through the needle or catheter. Early complications include perforations (vessels and other structures) that may be related to the needle, guidewire, dilator, or catheter, or later perforations related to catheter-induced erosion. There is great variability in human anatomy and it makes it difficult to cannulate the vein. Thus it is important to look with ultrasound before cannulation attempts to avoid puncture to the artery.

Ultrasonography is used increasingly at the bedside to assist in the placement of CVCs. Ultrasound is recommended for routine use in CVC placement; reports demonstrate that its use reduces complications in infants and children. Multiple Research Studies demonstrate decreased number of puncture attempts and lower complication rate (PTX, Hematoma) with ultrasound guidance. Some operators use the technology to mark the vein prior to attempted puncture (static technique), while others use real-time imaging to guide the needle puncture and CVC placement (dynamic technique). Higher success rates are generally found when real-time images are obtained and used to guide needle and catheter insertion during the procedure.

### Article No 18302

#### Comparing the prevalence of dysnatremia in 2 methods of IV maintenance fluid therapy with half saline versus standard 0.2 saline solutions in term neonates with sepsis

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**Objective:** Comparing the prevalence of dysnatremia in two methods of intravenous maintenance fluid therapy with half saline (75 mEq/L) versus 0.2 saline (30mEq/L) solutions in term neonates with sepsis.

**Methods:** In a double-blinded randomized clinical trial, Sixty term neonates (38-42 weeks of gestation) with sepsis were enrolled. Blood samples were taken with the aim of determining serum creatinine level, BUN, Na and K before the onset of treatment. Urine samples were taken to assess specific gravity and urinary output. Based on computerized random numerical table, the patients will be divided into two groups A and B. Group A, assigned to receive half-saline solution as maintenance intravenous fluid, and group B, assigned to receive the conventional 0.2 saline solution as maintenance. The above indicators were re-evaluated at 6, 24, and 48 hours after the initiation of treatment. The two groups were compared in respect to the incidence of dysnatremia, and other criteria such as urinary output and urinary specific gravity, BUN and Creatinine levels.

**Results:** No patient developed hyponatremia or hypernatremia after 48 hours of therapy. Sodium levels were significantly higher in half saline recipients 24 h ( $137.83 \pm 2.9$  vs  $134.4 \pm 1.92$  mmol/L), and 48h ( $137.67 \pm 1.99$  vs  $133.1 \pm 2.41$  mmol/L) after treatment ( $P < 0.001$ ). The urinary output, urine specific gravity, K, BUN and Creatinine levels were not significantly different in two groups.

**Conclusions:** The use of Half-Saline solution (75 mEq/L) as maintenance fluid did not increase the risk of hypernatremia after 48 hours when compared to 0.2 Saline solution, which is by now the conventional maintenance solution in neonates. Both half saline (Na 75 mEq/L or 0.45% NaCl) and Na 30 mEq/L NaCl or 0.2% solution can be used safely in term neonates.

## Article No 18303

## CKD registration in Iran

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## گزارشی از مطالعه مربوط به فاز اول سامانه ثبت آمار کودکان مبتلا به CKD در ایران

بیماری مزمن کلیه (Chronic Kidney Disease (CKD) از علل مهم ناخوشی و مرگ و میر در کودکان بویژه در کشورهای در حال توسعه می باشد. در کشور ما بعنوان یکی از ممالک در حال توسعه و با جامعه ۷۵ میلیونی (در سرشماری سال ۱۳۹۰) که ۲۱/۳۰۷/۶۵۰ نفر (۲۸/۴۱٪) از جمعیت آن را کودکان زیر ۱۸ سال تشکیل میدادند، اطلاعات جامعی در سطح ملی در رابطه با شیوع بیماری مزمن کلیه در جمعیت اطفال و نیز میزان بروز سالانه آن جمع آوری و به ثبت نرسیده بود. نتیجه اینکه ما هیچ اطلاع دقیقی از موارد جدید بیماری، شیوع و اتیولوژی بیماری مزمن کلیه در سنین مذکور و نیز هزینه هائی که از این بابت به سیستم بهداشتی کشور تحمیل می شد، در دست نداشتیم.

آگاهی از وسعت و اتیولوژی CKD در این جمعیت نه تنها در روشن نمودن گستردگی بیماری کمک کننده می باشد، بلکه بر اساس اطلاعات بدست آمده ضمن شناخت موانع و مشکلات احتمالی در زمینه تشخیص بیماری، میتوان با برنامه ریزی اصولی و ارائه راهکارهای مناسب اقدامات موثری را در رابطه با پیشگیری و با کاهش میزان ابتلاء به CKD در کودکان و درمان آنها معمول داشت. ما در فاز اول این بررسی از سال ۱۳۷۰ لغایت ۱۳۸۸، اکثریت کودکان مبتلا به CKD و ارجاع شده به کلینیک های نفرولوژی کودکان و بیمارستان های کودکان دارای بخش های نفرولوژی در تهران و مراکز سایر استانها را، با همکاری جمع کثیری از استادان نفرولوژیست در طب کودکان و شاغل در دانشگاه های علوم پزشکی سراسر کشور، مورد مطالعه قرار دادیم. در این مطالعه ۱۲۴۷ کودک مبتلا به CKD شامل ۶۶۲ پسر و ۵۸۵ دختر در محدوده سنی ۳ ماه تا ۱۸ سال با میانگین سنی ۴/۷۲ ± ۷/۶۹ مورد بررسی قرار گرفتند.

این میانگین در گروه همودیالیز ۳/۱۸ ± ۷/۷۹ سال، در گروه CAPD ۱/۸۶ ± ۴/۲۴ سال و در گروه پیوند کلیه ۱/۹۵ ± ۳/۴ سال بود.

شیوع CKD در کودکان مورد مطالعه از سال ۱۳۷۰ لغایت ۱۳۸۸ ابتدا از یک سیر صعودی و در سال های پایانی مطالعه از یک سیر نزولی برخوردار بود. میانگین بروز و شیوع سالانه CKD در Stage های ۲-۴ به ترتیب ۳/۳۴ و ۲۱/۹۵ مورد به ازای هر یک میلیون جمعیت اطفال (p.m.a.r.p) بود. میانگین بروز سالانه CKD در Stage های ۲-۴ و Stage ۵، به ترتیب ۰/۸۳ و ۲/۵۳ مورد به ازای هر یک میلیون جمعیت اطفال (p.m.a.r.p) گزارش گردید. همچنین میانگین شیوع سالانه CKD در Stage های ۲-۴ و Stage ۵ به ترتیب ۲/۹۸ و ۱۸/۹۸ مورد به ازای هر یک میلیون جمعیت اطفال (p.m.a.r.p) بود. میانگین پیگیری بیماران در محدوده ای بین ۴/۶۵ ± ۷/۱۹ سال قرار داشت. در مجموع میزان بقاء ۱، ۵ و ۱۰ ساله بیماران بترتیب ۹۸/۳٪، ۹۰/۷٪ و ۸۴/۸٪ گزارش شد.

آنومالی های مادرزادی کلیه و سیستم ادراری (CAKUT) در ۴۰/۰۱٪، گلومرولوپاتی ها در ۱۹٪ و علل ناشناخته در ۱۸/۲۸٪ و بیماری های کیستیک، ارثی و مادرزادی کلیه در ۱۱/۱۴٪ بیماران، از علل عمده CKD در کودکان مورد مطالعه بود.

سی و صد و ده کودک (۲۴/۸۵٪) روی درمان نگهدارنده، ۵۳۷ بیمار (۴۳/۰۶٪) تحت همودیالیز و در ۱۸۲ بیمار (۱۴/۵۹٪) از روش CAPD برای درمان بیماران استفاده شد. انجام پیوند کلیه در ۲۱۸ بیمار (۱۷/۴۸٪) از جمله یافته های گزارش شده در این مطالعه بود. مرگ و میر در ۱۳۸ مورد (۱۱/۰۶٪) از بیماران مشاهده شد.

نتیجه: بروز و شیوع CKD در کودکان و نوجوانان در ایران بطور نسبی کمتر از موارد گزارش شده در کشورهای اروپائی و سایر مطالعات مشابه می باشد. همانند بررسی های انجام شده در اروپا، CAKUT علت اصلی CKD در بیماران مورد مطالعه بود. اقداماتی از قبیل غربالگری با استفاده از اولتراسوند برای تشخیص CAKUT، درمان و پیگیری مداوم، می تواند در بهبود پیامد این گروه از کودکان کمک کننده باشد.

در هر حال علت CKD در بخش عمده ای از کودکان (۱۸/۳٪) ناشناخته بود که انجام تحقیقات بیشتر برای شناسایی فاکتور های مرتبط با پیشرفت بیماری بطرف CKD را ضروری می سازد. نکته قابل توجه در این مطالعه بالا بودن میزان بقاء بیماران نسبت به مطالعات مشابه بود.  
\*p.m.a.r.p : per million age-related population

### Article No 18305

#### Dipstick urinalysis screening of healthy neonates

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**Background:** Renal disease may accidentally be discovered during urinalysis. This study was conducted to examine the usefulness of dipstick urinalysis screening in healthy neonates for the diagnosis of underlying renal disease and to study the magnitude of abnormal urinalysis in apparently healthy neonates.

**Methods :** In this descriptive study, voided urine samples were obtained from 400 apparently healthy neonates and tested using urine dipstick. The reaction of dipstick strip was read visually by a trained nurse. In cases with an abnormal urine analysis, a second screen test was performed within a week, and for those with persistent abnormalities, complete diagnostic tests were done.

**Results :** On the first urinalysis, 375 (94%) subjects were normal and 25 (6%) had abnormalities: 23 had proteinuria (5.75%), one was blood positive (0.25%), and one was both protein and blood positive (0.25%). Male neonates had a higher proportion of proteinuria than female neonates ( $p=0.038$ ). In the second examination, proteinuria was found in five (1.25%) neonates, but the proportion of other abnormalities did not change. In follow-up investigations, ureteropelvic junction obstruction and vesicoureteral reflux were recognized in two infants who had blood-positive or combined blood- and protein-positive results on their first tests.

**Conclusion:** The findings of this study show that dipstick test during neonatal period could be used for early diagnosis of renal diseases.

**Key Words:** Dipstick, healthy neonate, urinalysis

### Article No 18307

#### Prevalence of hyponatremia among patients with acute urinary tract infection (UTI) in Bandar Abbas Pediatric Hospital during 2012-15

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**Introduction:** Urinary tract infection (UTI) is among the most prevalent bacterial infections among children. There is scarce research on the correlation of UTI and the emergence of hyponatremia. The present research aimed to investigate the prevalence of hyponatremia among patients with an acute UTI.

**Methods:** The present research is of a cross-sectional and retrospective in type and was conducted on 1-month to 15 year-old child patients suffering from an acute UTI hospitalized in Bandar Abbas Pediatric Hospital within 2012-15. The sample size was 1,096 and the data were collected from the patients' medical records. The subjects were divided into two groups: with or without hyponatremia. The data were recorded in terms of the existence of upper and lower urinary infection, lab parameters and DMSA scan results as well as renal anomalies. The data were finally analyzed via SPSS ver.16 through Mann-

Whitney U-test, Chi-squared test and Fisher's exact test. The level of significance was set at  $p < .05$ . Results: 71 subjects (6.5%) were afflicted with a reduced serum level of sodium. In the hyponatremia group, ESR, CRP and WBC levels in blood showed a statistically significant increase ( $p = .001$ ,  $p = .001$ ,  $p = .006$ ). The two groups showed to be significantly divergent in terms of the presence of kidney photopenic area and upper urinary infection ( $p = .016$ ,  $p = .043$ ).

**Discussion:** The present findings revealed that hyponatremia can be a symptom of urinary tract inflammation. It is also correlated with an inflammation degree in children with UTI. ESR, CRP and WBC levels are negatively correlated with the serum level of sodium. In patients with a kidney photopenic area the emergence rate of hyponatremia is higher.

**Conclusion:** Hyponatremia and high levels of CRP can be considered as two independent factors involved in predicting the emergence of renal damage in DMSA scan.

### Article No 18309

#### Changes of antibiotic pattern in Iranian children with urinary tract infection

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**Introduction:** Knowledge of local antimicrobial resistance patterns is essential for evidence-based empirical antibiotic prescribing. Resistance patterns to antimicrobial agents vary according to the geographic region; consequently, the choice of antimicrobial empiric treatment should not only be based on the most likely urinary tract pathogens but also on their updated local resistance patterns. Materials and

**Methods:** A retrospective analysis of the antimicrobial resistance within inpatient children with the positive urine isolates and final diagnosis of UTI over the 12 year period, 2005 to 2017, in Ali Asghar Children's Hospital was performed.

**Results:** In total, 958 female and 349 male positive cultures were analysed. E. coli (77.6%) was the most common causative agent of UTI in children. The overall resistance rates were as follows: Ampicillin (78%), Cefazolin (60%), Cotrimoxazole (57%), Cephalexin (54%), Nalidixic Acid (44%), Ceftriaxon (38%), Gentamycin (22%), Ciprofloxacin (18.6%), Cefepime (18.1%), Imipenem (12%), Amikacin (10%), Nitrofurantoin (8%). E. coli Resistance rate increasing significantly over the 2008 to 2011 although our study detected the least E. coli Resistance rate over 2015 to 2017. Higher antibiotic resistance rates were identified in the male population with the exception of cotrimoxazole. Overall resistance to Cotrimoxazole was related to age: 51% below and 61% above 1 year ( $p < 0.001$ ) and Resistance rate were higher to Amikacin, Gentamycin, Nitrofurantoin in (below 1 year old) children. Higher antibiotic resistance rates were identified in patient with anatomical abnormality and there is significantly association between Positive Antibiotic history in the past two weeks and higher resistance rate among Cephalosporins, Ampicillin, Cotrimoxazole and Nalidixic Acid.

**Conclusion:** anatomical abnormality, previous Antibiotic History, Age and Gender were among the risk factors associated with antimicrobial resistance. Statistical analysis of the resistance pattern trend during 12 years indicated the significant decrease in E. coli antibiotic resistance in the last three years. Decisions about empirical therapy for UTI are best made on a case-by-case basis based upon the demographic and clinical risk factors for higher resistance rates.

**Key words:** Urinary Tract Infections, Antimicrobial Drug Resistance, Pediatrics, Ali Asghar Children's Hospital

**Article No 18312****Peritoneal dialysis: Use beyond kidney failure**

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Peritoneal dialysis is one of the method of renal replacement therapy, which is known as a less invasive and more cost-effective than other types of dialysis. In recent decades, this method has been used for non-renal indications. Including acute pancreatitis, psoriasis, congestive heart failure, hypothermia and poisoning.

Unlike initial studies, recent research did not provide clear clinical benefits in using peritoneal dialysis for the treatment of acute pancreatitis and psoriasis, and by developing more effective treatment for these diseases, peritoneal dialysis is not currently indicated for them and is known to be one their historical therapies. However in the case of congestive heart failure, hypothermia and poisoning, it is an effective method.

The benefits of peritoneal dialysis for the treatment of systemic congestion in patients with heart failure have been shown in several studies and further studies are underway. Currently, one of the methods for ultrafiltration in patients with resistant heart failure is peritoneal dialysis.

One of the treatments of patients with severe hypothermia is core warming that can be achieved with peritoneal dialysis, especially in patients with unstable hemodynamics.

Another use of peritoneal dialysis is the removal of harmful substances or the administration of antidote in patients with poisoning. Currently, other studies have also been conducted to assess the benefits of peritoneal dialysis in other diseases that may be potential future indications of peritoneal dialysis including treatment of acute stroke and as an oxygenation modality.

**Article No 18313****Vitamin D Deficiency as an indicator of artificial light poisoning , A historic narrative view of vitamin d and human displacement**

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Life begins to evolve in the oceans over 1 billion years ago. Phytoplankton as an earlier habitant of earth took advantage of sunlight and used it as an energy source to generate carbohydrate and makes vitamin D. Role of vitamin D in this tiny factory of photosynthesis is protection of important components of cell, DNA and RNA and amino acids with double bonds. The capacity of vitamin D to absorb ultraviolet B in phototoxic wavelength of 290nm makes it a vital natural sunscreen for cells (1). Meanwhile this hormone is a photo chemical to signal organisms to move from surface to deep ocean in order to prevent excess exposure to photo toxic dosage of ultra violet B. (signal for movement & protection) (2).

All vertebrate need vitamin D for movement they make it from skin exposure to sun or consume precursors from sea staff that eat phytoplankton in food chain or from plants. (3)

Human and vitamin D

Vitamin d has had huge impact on evolution of human skin color .By gradual movement toward in higher latitude from equatorial area only females who have evolved to lighter skin were able to produce enough vitamin d, and have a normal pelvis anatomy to normally deliver a healthy baby; female with low vitamin d (probably dark skin with low vitamin ) were excluded from reproductive cycle due to maternal complications (4). This is the reason that residents of northern latitudes have lighter skin color now. Skin

pigmentation determined by one favoring photo protection near the equator and other favoring vitamin D production nearer the pole.

Two other histories from movement and vitamin D back to the industrial revolution. In 1600 majority of big cities in Europe hosted people who immigrated from rural to urban area, this movement accompanied by living in home close proximity to each other. Coal consumption made city heavily polluted, reports showed more than 80 % of children in these cities had rickets (5). In 1822 one century before Hess discovery of sunlight exposure as a treatment of rickets (1923), Sniadecki in Warsaw said lack of sun light is the reason of disease what was later accepted (6).

Force movement of black people is another history of movement and vitamin d. The slave trade took blacks out of Africa and settled them, among other places, across North America, Canada and Northern Europe where for months of the year sun rays strike less directly, drastically reducing the amount of sunlight which their skin were not adapted and was necessary to make adequate vitamins d. In that time medical report described a disease that is similar to hypocalcemia of vitamin d deficiency, it was called "black (7) diseases" that seen in black slave after a period of settling; with neurological symptoms and psychosis. The reason was fast displacement of black from one continent to other continent, it was slavery disease I called it. Through time, human adaptations to different solar beam have become more cultural than natural. Rapid human migrations with industrial revolution, increasing urbanization in modern life, and changes in lifestyle have created mismatches between skin pigmentation and environmental conditions (8). In the beginning of life adequate vitamin D was a signal for movement, force or rapid displacement in human history was accompanied with vitamin D deficiency.

Is it possible that vitamin d deficiency again be a sign of premature shift from one place to other place?

Or new type of slavery not limited to black but include all races black, white and yellow and not limited to one continent but pandemic. Vitamin d deficiency may be an alarm signal for abnormal mankind displacement from outdoor activities and sun exposure opportunity. It will be more important when we realize during sun exposure several other by product produce in skin like melatonin and tachestrol which have anti proliferative effect and regulate epidermal growth, in the other words vitamin d production is only 15% of by products that produce during solar exposure and other photo product has other benefit which will turn out in the future (9). If we have enough sun exposure in the environment and do not receive any vitamin D supplement, then adequate vitamin D (levels >30ng/dl) level will be a marker of adequacy of outdoor activity. Prescription of vitamin d to normalize 25 hydroxy vitamin D is a very simplistic approach to the problems that took place in our history for several times.

Let me leave you with a question what is the hazard of indoor activity? Evidences show indoor activity and lack of sun exposure is a risk for premature death like obesity, smoking and inactivity. It remains to reveal the impact of this behavior by huge human cost in future.

What do we use instead of natural sun light when we engage in indoor activity? Is it our new environmental hazard?



### Article No 18314

#### The relationship between low-birth weight and nephrotic syndrome in children

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**Introduction:** The body weight of an infant is one of the most important factors that influences its survival, growth and development. Many clinical studies have shown a higher risk of an aggravated course of renal disease in children born with low-birth weight (LBW), due to reduction in glomerular number and development. However, there are limited clinical studies about this relationship.

**Objectives:** This study was conducted to evaluate the relationship between LBW and risk of idiopathic nephrotic syndrome (INS) in children.

**Patients and Methods:** In this case-control study, we evaluated nephrotic syndrome patients who were referred to Imam Raza hospital. Seventy patients with nephrotic syndrome as case group and 140 healthy children under age 16 with normal birth weight as control group were enrolled in the study. Data including age, sex, and birth weight were collected from patient's records.

**Results:** This study revealed that the risk of nephrotic syndrome in patients with LBW is two times higher than those with normal birth weight. However, there was no statistically significant difference ( $\chi^2 = 1.58$ ,  $P = 0.12$ )

**Conclusion:** Although the result of this study cannot show a statistically significant relationship between INS and LBW, the risk of nephrotic syndrome was twice in the case group.

### Article No 18315

#### Intra-dialytic Hypotension in Hemodialysis Patients: A Single Center Evaluation

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**Introduction:** Intra-dialysis hypotension (IDH) occurs in 20- 55% of hemodialysis sessions and is more frequent among patients on long-term hemodialysis. We aimed to define the impacts of blood pressure status, inter-dialysis weight gain, vasodilators antihypertensive drugs, characteristics of hemodialysis, serum calcium, sodium, and albumin and hemoglobin concentrations on prevalence of intra-dialysis hypotension.

**Methods:** 44 hemodialysis cases in 508 dialysis sessions were evaluated for intra-dialysis hypotension. They included 19 girls and 25 boys aged 4.8- 25 years. A decrease in mean  $BP \geq 10$  mm Hg during hemodialysis was defined as hypotension.

**Results:** IDH was noted in 136 of 508(26%) dialysis sessions. It was significantly more prevalent in cases with normal systolic and diastolic blood pressures compared with those who had diastolic or systolic hypertension ( $P=0.014$  and  $P=0.005$  respectively). There were no meaningful differences in prevalence of hypotension based on characteristics and duration from onset of dialysis, and inter-dialysis weight gain ( $P>0.05$  for all). Also it was as common in cases treating with vasodilators drugs as those without ( $P=0.221$ ). Serum calcium, sodium and hemoglobin levels didn't have significant differences between cases with and without IDH, whereas those with IDH had a significantly lower serum albumin concentration ( $P=0.021$ ).

**Conclusion:** normotensive patients and cases with lower serum albumin concentrations are more prone to develop IDH. Measurement of blood pressure with shorter intervals in normotensive patients is recommended for early diagnosis of IDH.

**Key words:** Hemodialysis, blood pressure, IDH, dialysis sessions, serum albumin concentrating

### Article No 18316

#### Blood pressure of newborn with gestational age between 26-42 weeks in a tertiary center in Iran

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**Background and aim:** Blood pressure (BP) is a reflection of hemodynamic variables. It is an important vital sign and indicator of clinical stability. Accurate measurement and interpretation of this physiological signal is essential for the optimal management of the ill newborn. An increase in the awareness of

hypertension among neonates has resulted to increased ability to diagnose neonates with the disease. Objectives: This study aimed to determine BP values and percentiles in stable newborns in the first weeks of life and evaluate the relevant factors.

**Methods:** This prospective observational study was conducted on 320 term and preterm newborns with gestational age (GA) between 26 and 42 weeks between 2015-2017. Exclusion criteria included birth asphyxia; infants of mothers with hypertension, preeclampsia, gestational diabetes, type 1 diabetes mellitus or illicit substance use; major congenital anomaly. BP measurements were determined using the oscillometric technique with the neonate supine after an appropriate size cuff was applied on the right arm. Multichannel monitor was used to determine systolic BP (SBP), diastolic BP (DBP), and mean arterial pressure (MAP) by oscillometric method (ANSI/AAMI SP-10/2002 Memory 500 Records IBP 2 Channels (Up to 4)). Systolic and diastolic BPs were statistically analyzed by regression analysis for various percentiles (5<sup>th</sup> to 95<sup>th</sup>).

**Results:** This is made up of 185 (57.8 %) males and 135 (42.2 %) females with M: F ratio of 1.36:1. Birth weight ranged from 1650-4650 gr (mean = 2058.3 ± 852.5 gr). GA ranged from 26- 42 weeks (mean = 32.95 ± 3.97 weeks). Two hundred and twenty one (69.1 %) babies were delivered through cesarean section and 99 (30.9 %) deliveries were by vaginal delivery. Percentile charts (providing 5th, 10th, 25th, 50th, 75th, 95th values) have been developed. SBP, DBP and MAP showed a steady rise on the respective days that were comparable between different groups. Evidently, there were no statistically significant gender differences in mean body weight, gestational age and the BP parameters. Term babies were found to have higher SBP, DBP and MAP than their preterm counterparts on the respective days that were statistically highly significant. Neonates who were delivered vaginally had higher mean BP values for systolic, diastolic and mean than neonates delivered by cesarean section. There were no statistically significant difference in mean SBP, DBP and MAP recordings between males and females. However Female neonates had higher systolic BP values than male neonates.

**Conclusion:** The study provided normative BP values among neonates especially in the first 7 days of life. These values are recommended for use in the evaluation of BP in newborns. Data presented in this study mode specific BP percentile curves using an oscillometric method and serve as a valuable reference for physicians in dealing with the management of newborns in the neonatal unit.

### Article No 18317

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## Prevalence of hypertension and pre-hypertension in 7 – 11 Year old children in the city of Ahvaz in 2013

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**INTRODUCTION:** There are some studies about the prevalence of hypertension and prehypertension in different regions of Iran. Our study is unique in considering the prevalence of both hypertension and prehypertension in the South- West of Iran.

**MATERIAL AND METHODS:** This abstract is based on a graduation thesis of residency by Dr. Hosseini in 2013. It is a descriptive study involving 5% of the 7-11 year old population in Ahvaz . 5811 children were enrolled including 2907 girls and 2904 boys.

**RESULTS:** The prevalence of HTN and pre HTN among 7 to 11 year old children was 8.4% and 7.8% respectively. The prevalence of HTN in boys and girls was 8.6 and 8.2% respectively. The prevalence of pre HTN in boys and girls was 8.5 and 7.1% respectively. When considering the height percentile in children the prevalence of HTN and preHTN increased with the increase in height percentile in both boys and girls. With regard to BMI , the prevalence of HTN and preHTN increased in both boys and girls. In comparison to the most previous studies involving large numbers of children in Tehran and other cities , there seems to be an increased prevalence of HTN which can be ascribed to obesity epidemic , sedentary life style, and also geographic , nutritional, and genetic factors.

**CONCLUSION:** The prevalence of HTN and Pre HTN is increasing compared with previous years.  
**KEYWORDS:** hypertension, prehypertension, prevalence

### *Article No 18319*

#### **Mambranous GN. pathology review**

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Membranous nephropathy is a rare histologic entity in children, which usually presents as nephrotic syndrome or asymptomatic proteinuria. It is one of the most common causes of primary nephrotic syndrome in adults, it contributes to <5% of cases in children. It is characterized histologically by the uniform thickening of the glomerular capillary wall on light microscopy. This thickening is associated with subepithelial immune complex deposits that appear as granular deposits of immunoglobulin (Ig) G on immunofluorescence and as electron-dense deposits on electron microscopy.

In children, secondary causes of MN have been associated with conditions such as systemic lupus erythematosus (SLE), hepatitis B or C infection, secondary and congenital syphilis, malaria, and Epstein Barr Virus (EBV) infection. Other rare underlying causes are C4 deficiency, selective IgA deficiency, or antitubular basement membrane antibodies.

### *Article No 18322*

#### **Electrone Microscopy finding in Common Kidney Disease**

Fatemeh Nili

Tehran University of Medical Sciences, Tehran, Iran-

Electron microscopy is a part of routine pathologic assessment of kidney biopsies. Although it has a confirmatory role in many diseases, it is an essential tool for the diagnosis of some other disorders. In this brief review, we summarize some diagnostic ultrastructural findings in common glomerular diseases. Minimal change disease and focal and segmental glomerulosclerosis (FSGS) are podocytopathic disorders ultrastructurally diagnosed by extensive effacement of Visceral Foot Processes and absence of immune-type deposits. Distinction between unsampled FSGS and minimal change disease could be challenging on electron microscopy as the light microscopy.

For accurate staging of membranous glomerulopathy, electron microscopy is necessary. Presence of small sparse subepithelial deposits without GBM thickening is indicative of stage I. On stage II, GBM thickening and subepithelial spikes would be obvious. When the new basement membrane surrounds the deposits, stage III could be suggested, and finally stage IV shows extensive resorption of intramembranous deposits and foci of sclerosis.

Dense deposit disease is a type of C3 glomerulopathy which is characterized by deposition of mesangial and intramembranous extremely dense deposits. Electron microscopy is crucial to make the diagnosis. In problematic situations, presence of subepithelial humps are supportive of post-infectious glomerulonephritis.

Electron microscopy is necessary for the diagnosis of Alport syndrome and thin membrane disease (Benign familial hematuria). Irregular GBM with alternating foci of thickening, thinning and multilayering of lamina densa are diagnostic findings for Alport syndrome.

In some patients, widespread thinning of the GBM is the only pathologic finding, a situation that is reminiscent of thin basement membrane disease. So, thin GBM is a structural abnormality observed in

several progressive or benign nonprogressive hematuric disorders. Further investigations including IHC or molecular study for  $\alpha$  chains of collagen type IV are required in these patients.

Diagnosis of some other rare disorders such as Fabry disease, LCAT deficiency, Pierson syndrome, Collagen III glomerulopathy, Nail-patella syndrome and.... is demonstrated on Electron microscopy as well.

### *Article No 18324*

#### **Mucocutaneous Lymph Node Syndrome; new strategies**

Reza Shiari

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Kawasaki Disease (KD) is a self-limited vasculitis of unknown etiology that predominantly affects children younger than 5 years. It is now the most common cause of acquired heart disease in children in Asia. KD was first reported in 1967 by professor Tomisaku Kawasaki. He described the clinical signs and symptoms of 50 Japanese children with acute mucocutaneous lymph node syndrome. One year later, Yamamoto T, published the electrocardiogram abnormalities of KD patients. Finally, Japanese physicians established the relationship between KD and coronary vasculitis in 1972.

***Back to the Future: 1871–1984 and Other Riddles of KD:*** More than 100 years of pathologic records from the first description of fatal KD, by Samuel Gee in 1871, (Gee, St Barth Hosp Rep 7:148, 1871) “infantile periarteritis nodosa”.

***The Era of Enhanced Discovery and International Collaboration: 1984–2015:*** The most recent period, from the mid-1980s to the present, marks the period of international cooperation, heightened interest, and scientific progress in the understanding of this still enigmatic disease. Histopathological observation of Kawasaki disease has focused on the coronary artery because coronary arterial lesions are directly associated with mortality and long-term outcomes. However, noncardiac lesions must also be considered when describing KD pathology and etiology. ITPKC and CASP3 are common susceptibility genes for Kawasaki disease. The recent identification of the FCGR2A, BLK, CD40, and HLA class II gene regions in genome-wide association studies has shed new light on the pathogenesis of Kawasaki disease. Although the cause of KD remains unknown, understanding of its pathogenesis has increased. An overt immune reaction triggered by unknown infectious agents may cause systemic vasculitis. The mediators of this reaction are mainly inflammatory cytokines such as TNF- $\alpha$ , IL-1 $\beta$ , and IFN- $\gamma$ . The agent of KD remains unknown after more than 40 years of intensive research, but new information from analyses of KD time series from locations worldwide suggests that KD activity is modulated by weather and climate processes. Heavy metals such as zinc and mercury can be transported on aerosolized particles and act as haptens that render antigenic the proteins to which they bind.

The standard therapy for acute Kawasaki disease is high-dose IVIG, 2 g/kg, over 8–12 h, together with aspirin, for its anti-inflammatory and then anti-platelet effects. Despite such treatment, however, up to 5% of children will develop coronary artery aneurysms.

Adjunctive therapies have included additional IVIG therapy, corticosteroids, tumor necrosis factor (TNF)- $\alpha$  blockers (e.g., infliximab, etanercept), calcineurin inhibitors (e.g., cyclosporine), interleukin-1 receptor antagonist (IL1RA) agents (e.g., anakinra), MTX, and cyclophosphamide.

Among these adjunctive therapies, only corticosteroids have been proven to be effective in reducing the incidence of coronary artery aneurysms, in a phase III randomized trial in a high-risk Japanese population.

### *Article No 18328*

#### **Renal manifestations in children with familial Mediterranean Fever**

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Familial Mediterranean Fever (FMF) is an inherited condition consists of recurrent episodes of inflammation in the abdomen, chest, or joints often accompanied by fever and sometimes a rash or headache. The gene responsible for FMF, *MEFV*, encodes a protein that is expected to be a down regulator of inflammation.

The major renal involvement in FMF is the occurrence of amyloidosis that primarily affects the kidneys and primarily manifests as a nephropathy that passes through consecutive stages of proteinuria, nephrotic syndrome, uremia, end-stage renal failure and death. Association between FMF and non-amyloid glomerulopathies are unusual. It is estimated that about half of the renal involvement in FMF be non-amyloid. Other renal manifestations of FMF includes mesangial proliferative glomerulonephritis (MsPGN), rapid progressive glomerulonephritis (RPGN), focal segmental glomerulosclerosis (FSGS), membranoproliferative glomerulonephritis (MPGN).

We report 2 children who presented with recurrent episodes of fever and abdominal pain and positive test for *MEFV* gene mutations. One had heterozygous mutation at E148Q and had persistent proteinuria whose renal biopsy showed no deposition of amyloid proteins. Another case had heterozygous mutation at R761H and showed systemic hypertension, hematuria and Nutcracker Syndrome.

Our study suggests that in patients with FMF, non-amyloid renal involvements should be considered in the differential diagnosis in addition to amyloidosis.

### Article No 18329



## Establishment of Colonic Dialysis Model In Uremic Rats By Right Nephrectomy And Left Partial Nephrectomy.

Kajbafzadeh AM, Sabetkish N, Sabetkish S.

J Pediatr Urol. 2018 Feb 7. pii: S1477-5131(18)30027-5. doi: 10.1016/j.jpuro.2017.11.024. [Epub ahead of print]

### Article No 18330



## Pathologic Findings of Membranoproliferative Glomerulonephritis

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Membranoproliferative glomerulonephritis (MPGN) describes a pattern of glomerular injury with common histologic features of glomerular capillary wall thickening (membrano-) and hypercellularity in the glomerular tufts (-proliferative). MPGN was historically classified into three morphologic types: type I, with presence of immune deposits in the subendothelial and mesangial areas; type II, with electron-dense deposits within the basement membrane; and type III, with complex GBM formation with subendothelial and subepithelial electron-dense deposits that are bridged by intramembranous deposits. MPGN can be primary (or idiopathic, most commonly in children) or secondary to infectious and autoimmune disease. A unifying characteristic of all types of MPGN is hypocomplementemia (low C3). MPGN type II, known as

dense deposit disease (DDD), is now considered a separate entity from MPGN types I and III, since it has unique pathogenic and clinical features.

The disease is characterized by functional impairment of the glomerular basement membrane (GBM), causing progressive loss of renal function that eventually results in end-stage renal disease (ESRD). Clinical features at first manifestation are hematuria, proteinuria (nephrotic range), impaired renal function and hypertension

## Poster Presentation

### P1:

#### The Frequency of Complement Dysregulation in Hemolytic Uremic Syndrome Suspicious Cases- Two Centers Study

Hooman N<sup>1</sup>, Hosseini S.<sup>1</sup>, Otukesh H<sup>1</sup>, Hossieni shamsabadi R<sup>1</sup>, Dalirani R<sup>2</sup>, Esfandiar N<sup>2</sup>, Nikavar A<sup>1</sup>, Mohkam M<sup>2</sup>, Sharifian M<sup>2</sup>, Afshin A<sup>2</sup>, Haghighi B<sup>1</sup>, Tabtabee TH<sup>2</sup>, Afsharkhas L<sup>1</sup>.

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**Background and Aim:** Hemolytic uremic syndrome (HUS) consists of heterogeneous group of diseases which share same clinical and pathological manifestation. Complement consumption and dysregulation is one of the features in all subtypes of HUS. Classic and some inhibitory factors in the alternative pathway are usually measured in order to discriminate the subtype of HUS. We do not know how much these measurements were useful in our setting. The aim of this study is to find the percentage of abnormal level of complement in the assessment of underlying disease of HUS.

**Materials and Methods:** This was a descriptive retrospective conducted between 2005 and 2017. Data of all children aged less than 18 years old suspicious to HUS and admitted in Ali-asghar children hospital or Mofid hospital were reviewed. Complement levels (C3, C4, CH50), inhibitory factors (H, I, B), CD46, ADAMTS 13, ADAMTS 13 AB were recorded and the laboratory cut point used to classify the complements to low, normal or high level.

**Results:** 76 patients (36 females, 40 males) with mean age of 5.2 yrs (3 m -24 yrs) were analyzed. Diarrhea positive HUS in 6 (7.8%), pneumococcal HUS in 1 (1.3%), TTP in 7 (9.1) and 37 patient with atypical HUS were diagnosed. The Most common clinical manifestation at the admission time was non bloody diarrhea (12.7%).

The frequency of complement deregulation was shown in table 1. It should be considered that some of the patients had received FFP prior to check serum complement level. Patients with both factor H and I deficient consist 2.5% and 2.5% had low levels of all factors H, I, B.

During the last 12 years, 25% of cases achieved full recovery, 5.2% died, and the rest are on renal replacement therapy.

**Conclusion:** This study revealed that the routine panel of HUS is diagnostic in about 10% and expanding the diagnostic tests in addition genetic study for underlying disease is mandatory.

Table 1- The frequency of complement dysregulation according Plasma Level

	Decreased	elevated	normal Values
C3	43%		43%
C4	14%	10.4%	53.2%
CH50	20.8%	3.8%	46.8
Factor H	9.1%	16.9%	68.8%
Factor I	37.7%	11.7%	44.2%
Factor B	5.2%	20.8%	67.5%
ADAMTS 13	6.5%	7.8%	72.7%
ADAMTS13 AB	-	16.5%	74%
CD46	9.1%	-	70%

**P 2:**

**The volume of plasma to achieve remission in atypical hemolytic uremic syndrome**

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Iran University of Medical Sciences, Tehran, Iran

Background and aim: Plasma infusion or plasma exchange is the rescue therapy for familial or recurrent hemolytic uremic syndrome. The recommended volume of plasma infusion is 20-40ml/kg. There is no document to show how much of plasma volume actually get the patient to remission. Material and methods: This was an observational retrospective study between March 2007 and 2015. All patients with diagnosis of recurrent or familial atypical hemolytic uremic syndrome (aHUS) admitted in Ali-Asgar Children Hospital included. The total volume of plasma required to normalize platelet, LDH, eliminate hemolysis, and decrease serum creatinin was calculate. Patients with TTP, vasculitis, and postinfectious were excluded. Results: Between 2007 and 2015, twenty-four patients (M-13, F-11) with diagnosis of aHUS were included. Two patents were excluded, one occurred after sepsis and the other did not receive plasma infusion. The median age (range) was 53 months (3-144 m). 20 patients achieved remission by plasma infusion (under PD-5, under HD-4) and two patients treated by plasmapheresis. The median (range) total plasma volume required for remission was 99ml/kg (43-235 ml/kg). Conclusion: This study showed recurrent or familial HUS in acute phase require at least twice total plasma volume than those volume recommended in literature in order to control the relapse phase of disease.

**P3:**

**Rituximab dependent Nephrotic Syndrome (A case report )**

Ali Derakhsahn , Mitra Basiratnia, Mohammad Hossein Fallahzadeh

Shiraz University of Medical Sciences, Shiraz, Iran

**Introduction:** Poor prognosis is anticipated in children with FSGS despite various treatment options. Herein we report a case with good prognosis although very expensive and still challenging regarding the unknown long term side effects of Rituximab.

**Case report**

A 34-month old girl presented with features of nephrotic syndrome on 22/02/2010. She was resistant to a classic course of 60mg/m<sup>2</sup> daily prednisolone. A kidney biopsy revealed tip variant FSGS with 5%interstitialfibrosis.Two doses of Rituximab, 375/m<sup>2</sup>, was prescribedas well as tacrolimus 0.5mg q12h which was added to her low dose alternate day prednisolone. It took about a year for her to respond completely. Six months later she had a relapse which responded to increasing the dose of tacrolimus within 3 months, another relapse occurred 4 months later with a tacrolimus level of 7.8ng/ml; therefore, 2 more doses of Rituximab were prescribed on 25/09/2012. Since this date until her last relapse on 25/10/2017 sheexperienced 6 more relapses, with 10-13kg weight gain and a response to 2 doses of Rituximab within 2months on each relapse. Meanwhile she was on enalapril, spironolactone,low dose prednisolone and tacrolimus. Until now she has received 16 doses of rituximab.

Conclusion: This was a challenging case of rituximab dependent nephrotic syndrome (if we can use this term for the first time) and even more challenging is the frequent doses of rituximab with unknown untoward long term side effects.

Key words: nephrotic syndrome, FSGS, rituximab dependent

#### P 4:

##### **Rituximab efficacy on complicated refractory nephrotic syndrome: case report**

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This is a 16-year-old girl presentation who involved nephrotic syndrome (NS) since 11 years ago with biopsy exhibited minimal changed NS. Her full blunt nephrotic presentation was accompanied with normal GFR, no hematuria and without skin rash or hypertension at onset. Her serum complement assays were within normal range and immunofluorescence renal histology was negative. Although initial treatment with oral prednisolone was effective, but complicated by refractory and relapsing course and developed adverse drug toxicity such as cushingoid appearance, hypertension, short stature and low bone density. She received multiple adjuvant therapy with levamisole, Azathioprine, cyclophosphamide, cyclosporine and mycophenolate mofetil. Although Tacrolimus have had dramatic response, finally proteinuria reappear and sever skin rash proven biopsy pustular psoriasis developed. Rituximab (Zytux™ Aryogen) was started instead of tacrolimus, 4 weekly doses followed by one booster at sixth month. Her proteinuria diminished since third week of treatment and she experienced no relapse at one year follow-up with resolved skin rash. These observation suggest the efficacy of rituximab in patients with difficult nephrotic syndromewho had either responded unsatisfactorily to treatment with multiple immunosuppressive agents or had features indicating of medication-related toxicity. Persistent proteinuria known as poor prognostic predictor regarding renal function in nephrotic syndrome and the use of effective and safe medication such as rituximab in patients with refractory and complicated nephrotic syndrome is highly recommended.

**Keywords:** nephrotic syndrome, steroid resistance, rituximab, children

#### P 5:

##### **Dysurian as first Presentation of Bladder Lymphoma**

Mohamad Reza Razavi<sup>1</sup>, Khadijeh Rafsanjani Arjmandi<sup>2</sup>, Seyed Mojtaba Musavai Khoshdel<sup>2</sup>, Mohamad Molavi<sup>2</sup>, Mahtab Rahbar<sup>2</sup>, Nakysa Hooman<sup>2</sup>

<sup>1</sup> Qom University of Medical Science, Tehran, Iran, <sup>2</sup> Iran University of Medical Sciences, Tehran, Iran

A 5-year old boy presented with dysuria and bright red colored urine two days ago. Physical exam was remarkable for sever cachexia, and suprapubic tenderness. In primary study BUN and creatinin were normal but urine analysis had puyria and RBC, renal sonogram reported increment of bladder thickness. With suspicious to cystitis antibiotic was started, subsequently symptoms relieved. Two weeks later, he cam with hematuria and abdominal pain. A mass in supra pubic area was palpable. Foley catheter was inserted and the small amount of urine was drained. Repeated renal scan confirmed a bladder thickness of 28 mm with bilateral hydronephrosis. BUN and creatinin elevated and uric acid level was 23. Patient became oliguric. Abdominal CT scan without contrast revealed bladder thickness and bilateral hydronephrosis predominantly in left side ( figure one) . with diagnosis of post renal acute kidney injury

cystoscopy was done . Left uretero vesico junction was completely obstructed and the attempts for setting JJ in two side was unsuccessful and just biopsy was done. Tube nephrostomy helped to drain urine and creatinin decreased. Bone marrow aspiration and bladder biopsy( figure two) were in favor of leukemic lymphoma. Bone marrow replacement by large lymphoblast with prominent cytoplasmic vacuolation (Burkitt lymphoma/ ALL L3). Immunophenotype was increased (CD10, CD20, CD45, and HLA-DR). and SIgM was also increased. Standard protocol of chemotherapy was started (LMB89).

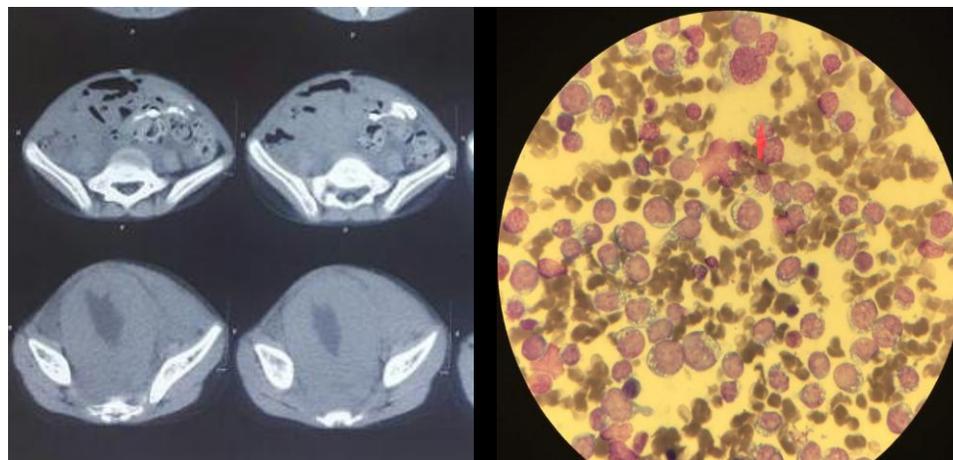
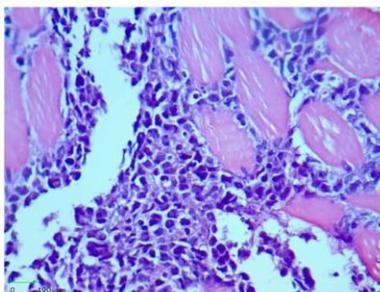
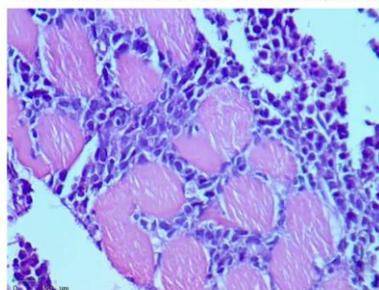


Fig.1- Abdominal CT scan showed bladder thickness( Left), Figure 2- bone marrow aspiration in favor of lymphoma ( Right)

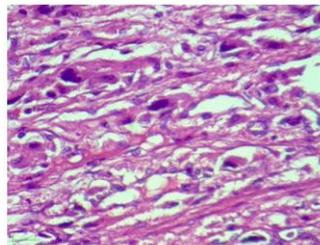
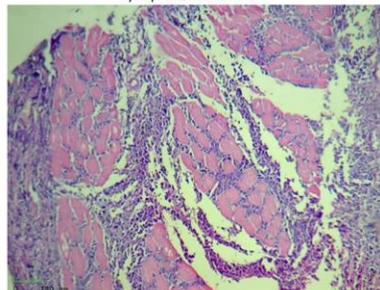
Muscular layer of urinary bladder infiltrated with lymphoblastic cell



Muscular layer of urinary bladder infiltrated with lymphoblastic cell



Muscular layer of urinary bladder infiltrated with lymphoblastic cell



**P 6:****A patient with pheochromocytoma and paraganglioma in different presentation times**

Davoud Amirkashani , Nakysa Hooman , Mahtab Rahbar , Seyed Mojtba Musavai Khoshdel

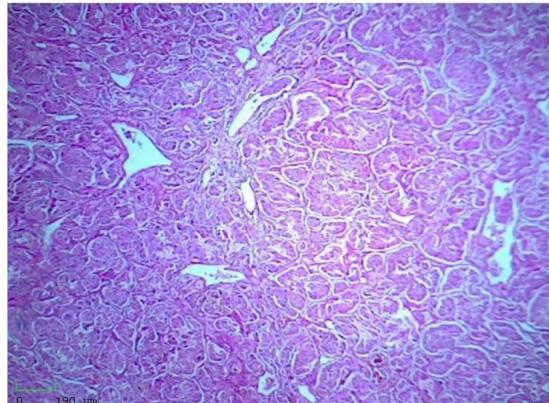
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The patient is a 15 years old boy admitted for excision of extra adrenal tumor. He complained from intermittent headache and palpitation and diaphoresis. Symptoms have been present from almost a few months before than refer to hospital. Blood pressure was measured and mild hypertension was detected. Therefore holter monitoring also was done and had been proven that patient was involved by persistent hypertension.

According to previous history , patient was investigated for 24-h collective catecholamins of urine (neumetanephrine, metanephrine, neurepinephrin , epinephrine, VMA and HWA). The patient had been underwent open adrenalectomy for pheochromocytoma almost last 3 years ago. In recent admission, after detection of rising urine catecholamins more than 4 times upper limit of normal range. Immediately, imaging was done and extra adrenal tumor was detected. After 10 days control of blood pressure by alfa blocker, laparoscopic excision of tumor was done.

Hypertension and other symptoms subsided and antihypertensive drugs discontinued. Pathologic study was shown nesting (zellballen), polygonal, spindle shaped cells in rich vascular network due to extra adrenal paraganglioma. It's interesting that genetic results about RET was undetected and other mutations are under investigating.

Zellballen (small nests or alveolar pattern), trabecular or solid patterns of polygonal / spindle shaped cells in rich vascular network

**P 7:****Spontaneous remission in a child with new onset NS . following ARF**

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**Introduction :** Either transient or permanent remission in NS is a well-known event. There are very old reports of spontaneous remission with measles and also a case of steroid resistant NS with Varicella infection. We report a case of nephrotic syndrome who developed acute renal failure (ARF) during his initial presentation and when his renal function recovered he was in remission. Spontaneous remission of NS with ARF has not been described in the written literature so far.

A 6.5-year-old boy presented with acute onset of generalized edema, hypo-albuminemia (Alb. 2.1 mg/dl), Hypercholesterolemia (Cho. 345 mg/dl), ESR 61 mm/hr, BUN 28, Cr 0.6 mg/dl, urine protein/Cr ratio

8. Meanwhile he developed gastroenteritis and dehydration. One week following his initial presentation, he was referred to our center with severe oliguria and his lab data was as follow: BUN 95 mg/dl, Cr 7.1 mg/dl Na and K 134 and 6.4 respectively. Four sessions of alternateday Hemodialysis (HD) was performed. His urine out-put showed a gradual increase and his renal function recovered completely within 2 weeks. The striking feature in this particular case was the remission of nephrotic syndrome with the recovery of renal function. After about 21 months he is still in remission.

**Conclusion:** Permanent remission of nephrotic syndrome following acute renal failure is described for the first time in the literature.

**Key words:** Nephrotic syndrome, spontaneous remission, acute renal failure, permanent

## P 8:

### Moyamoya syndrome in a child with Schimke immuno-osseous dysplasia

Mitra Basiratnia

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**Introduction:** Schimke Immuno-Osseous Dysplasia (SIOD) is a rare autosomal recessive disease caused by a biallelic mutation in SMARCAL1 gene. Typical findings in Schimke Immuno-Osseous Dysplasia include spondylo-epiphyseal dysplasia, steroid resistance nephrotic syndrome, progressive renal failure, T-cell immunodeficiency, bone marrow failure, and cerebral infarction. Herein, we describe a 9-year-old girl with SIOD.

**Case report:** The proband was the third offspring of healthy consanguineous parents (cousins). She presented with failure to thrive in infancy. At the age of 4 years nephrotic syndrome was confirmed for her. At the age of 6 years she developed right upper extremity paresthesia and weakness and brain MRI showed ischemic and hemorrhagic infarct in the left parieto-occipital and left caudate lobe. Two years later, she developed slurred speech and paresthesia, and weakness was progressed to her lower extremities. Brain MRI revealed new acute ischemic infarction in the right temporoparietal lobe. Encephalomalacia with surrounding gliosis was also noted involving the left parieto-occipital lobe because of the old infarction. Complete obstruction of the right MCA from the proximal part was seen in brain MRA. There was also evidence of significant wall irregularity and stenosis in basilar artery, PCA, ACA, internal carotid arteries, and vertebral arteries. These findings were in favour of moyamoya syndrome. In the last admission at the age of 9 years, she had new cerebral ischemia, developed seizure, and finally died because of cardiopulmonary arrest following pulmonary hemorrhage. Analysis of SMARCAL1 gene [1] revealed a homozygous nonsynonymous homozygous mutation c. [2459G>A].

**Conclusion:** Moyamoya syndrome should be considered as a diagnosis in a child with SIOD and cerebral infarction.

## P 9:

### NPHS2 gene in steroid-resistant nephrotic syndrome: prevalence, clinical course, and mutational spectrum in South-West Iranian children

Mitra Basiratnia

Shiraz University of Medical Sciences, Shiraz, Iran

**Introduction:** Mutations in podocin (NPHS2) gene have the key role in the pathogenesis of steroid-resistant nephrotic syndrome (SRNS) in children, but data is scarce regarding their prevalence and natural course among different all ethnic groups. This study was aimed to demonstrate the spectrum of NPHS2 mutations in children with SRNS and to compare the clinical course of disease in patients with and without mutation.

**Materials and Methods:** All 8 exons of NPHS2 were sequenced in 99 children, including 49 with SRNS and 50 with steroid-sensitive nephrotic syndrome (control group) by DNA sequencing.

**Results:** The prevalence rates of NPHS2 gene mutation among children with SRNS and SSNS were 31% and 4%, respectively. The prevalence rates of mutation among familial and sporadic forms were 57% and 26%, respectively. Thirty-three percent of the children experienced recurrence of primary disease after kidney transplantation, none of whom had a mutation. The clinical response to treatment was poorer in children with mutation in comparison with patients without mutation (12% versus 32%, respectively; odds ratio, 3.29, 95% confidence interval, 0.40 to 25.64). Patients with and without mutation could not be differentiated by demographic and histological features, glomerular filtration rate at onset, hypertension, progression to end-stage renal disease, and proteinuria.

**Conclusions:** Mutations of NPHS2 gene are frequent among Iranian children with SRNS. Regarding similar clinical features in patients with and without mutation and poor response to pharmacotherapy in patients with mutation, a molecular approach might be necessary for different treatment plans and prediction of prognosis.

### P10:

#### Comparison of the efficacy and safety between Pdpoinetin and Cinapoinetin in the Remodeling of Anemia in Children with Chronic kidney disease-Study Protocol

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**Introduction:** Chronic Kidney Disease (CKD) in children is accompanied with high mortality and morbidity. Anemia is one of the most common of these complications. Since the Erythropoietin deficiency is the main cause of anemia, then the use of Epoietin is the principal method for treatment of anemia in these patients. with consideration that two different Epoietin (Pdpoinetin with alpha chain & Cinapoinetin with beta chain) are available for use in treatment, We designed an study that compare efficacy and safety of these two drugs in children with CKD and anemia.

**Study Aim:** Comparison of the effectiveness and safety between Alpha chain Erythropoietin and Beta chain Erythropoietin in the Remodeling of Anemia in Children with Chronic Renal Failure

**Methods:** This study is randomized clinical trial that will perform on children with anemia due to CKD that refer to Aliasghar Children Hospital in Tehran during 2017-2018.

In this clinical trial (phase IV), the 40 patients are divided in two parallel groups (each group 20 patients) according to simple randomization based on table of random numbers:

- 1) Alpha chain erythropoietin (PDpoinetin, Pooyesh Darou co).
- 2) Beta chain erythropoietin (Cinapoinetin, Cinnagen co).

In these two groups, the drugs are prescribed based on KDIGO guideline with initiation dose of 100 unit/Kg/week for new patients and same previous dose in old patients. The route of prescription is SQ. The patients informations record every months, for three months and in the end of study, we will compare the efficacy and safety of these drugs in remodeling of anemia in CKD.

**Result:** It is an ongoing

**Ethical Cod:** IR.IUMS.FMD.REC 1396.9511359001

Trial ID: 29868

### P 11:

#### Investigation of urine random calcium, uric acid and oxalate urinary system ultrasonography findings in 7-11 year old students: A multicenter study

Elham Pournabakhtyaran, Masoumeh Mohkam, Abdollah Karimi, Mohsen Akhavan- Sepahi  
Pediatric Infections Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran

**Background and aim:** Renal diseases can be asymptomatic even in progressive disorders, so detecting urine and ultra sonographic abnormalities may help early diagnosis and prevention of disease occurrence. We aimed to investigate random urine parameters and urinary system ultrasonography findings in 7-11 year old students.

**Methods:** Healthy students from Tehran and Qom, cities of Iran were enrolled in a prospective descriptive study. We evaluated students by sex, age, weight, height, BMI. Then, they were examined by a fresh clean urine sample and ultrasonography of urinary tract. Urine was investigated for urine Ca/Cr, urine Oxalate/Cr, urine Citrate/Cr.

**Results:** From 932 students, 47.9% were female and 52.1% were male. The age range was 7-11 year and mean age was 9.08 year. History of renal disease and UTI were seen in 1.1% and 9.9% of students, respectively. In 78% of students ultrasonography was normal and in 22% it was abnormal. Abnormal findings included, Hydro nephrosis in 1.1%, fullness in 0.1%, duplication in 3%, stone in 0.7%, decrease size of kidneys in 0.4%, increased bladder thickness in 8.9% and other abnormal findings in 7.8%. Abnormal urine findings included, urine Ca/Cr in 10.9%, urine Oxalate/Cr in 12.8%, urine Uric Acid/Cr in 5.4%, urine Citrate/Cr in 96.9%.

**Conclusion:** According to the results, it is beneficial to perform these urinary screen tests to prevent progression to urinary system diseases.

**Key words:** urine analysis, ultrasonography, Urolithiases

### P 12:

#### Pure Red Cell Anemia a Complication of Recombinant Erythropoietin

Tahere Heydari, Nakysa Hooman , Gholamreza Bahoush  
Iran University of Medical Sciences, Tehran, Iran

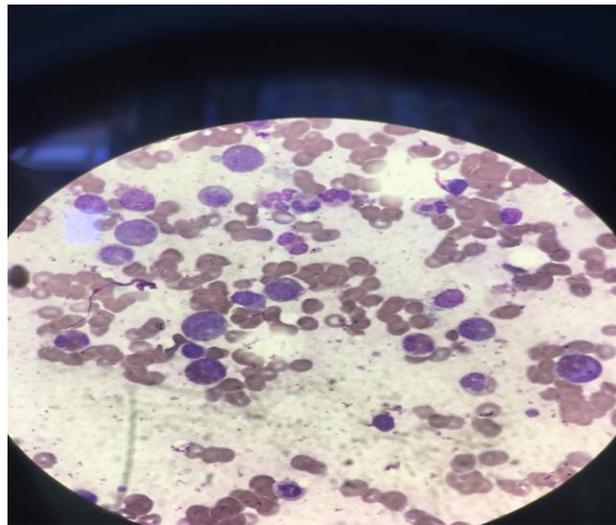
**Recombinant erythropoietin (EPO) is used to treat anemia in chronic kidney disease (CKD) to avoid multiple transfusions. Pure red cell aplasia (PRCA) is rare serious complication of EPO. The incidence increased considerably in 1998 decreased sharply since 2003.**

Hereby we presented a 5 -year old boy admitted with severe anemia. The underlying disease was nephritic syndrome with a progressive course that led to end stage renal disease. He went on CAPD with 600/m<sup>2</sup> three four times per day of dextrose 1.45% fluid and 100 ml extraneal at night since 2016. The peritoneal clearance and PET were checked every three months. The baseline hemoglobin (Hb) was 8.9 g/d and EPO-alpha ( PD-poetin) started with dosage of 300 U/kg/week. The Hb gradually increased to 11.6 g/dl and was persistently stable. After being 19 months on CAPD, the Hb declined to 6 g/dl, enalapril discontinued, EPO and PD dosages increased. Subsequently, the patient admitted with malaise and poor feeding the next month, Hb was 3 g/dl. The trough evaluation for the cause of severe anemia was done. Bone marrow aspiration revealed pure red cell aplasia .

With diagnosis of EPO induced PRCA, EPO stopped, transfusion was started, prednisolon 2mg/kg/day and IVIG 400mg/m<sup>2</sup> and then every two weeks while prednisolon tapered off. The Hb improved to 6 g/dl and the need to transfusion declined. He prepared for renal transplantation one month later. Kidney transplantation was unsuccessful and he passed away by massive hemorrhage and cardiac arrest.

Conclusion: In the case of severe anemia in CKD child full evaluation for detecting underlying etiology is important

Keywords: *Pure red cell aplasia, Anti-erythropoietin antibodies, Immunosuppressant therapy, Dialysis, Erythropoietin*



### P13:

#### Resistance to Erythropoietin-Stimulating Agents in Chronic Kidney Disease Children - Case Series

Neda Tavakoli , Nakysa Hooman

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**Background & aim:** Routine clinical and laboratory assessments facilitate early diagnosis of erythropoietin (EPO) unresponsiveness due to non adherence, hyperparathyroidism, malnutrition, inadequate dosages, some medication or resistant. The aim of this study was to find the frequency and the cause of EPO resistant in CKD children.

**Method:** This was an observational retrospective study. The lab tests of CKD children on EPO between 2008 and 2017 were reviewed. Hemoglobin less than 10mg/dl considered as resistant. Demographic data, EPO dosage, medications, underlying disease, the mode of dialysis and lab tests collected.

**Result:** Among 20 cases with CKD , 8(40%) had Hb<10 mg/dl. Male to female ratio was 1.66. The median range of age was 7.9 years old and of Hb was 6.25 mg/dl. (table 1) . Three cases(37.5%) had true resistant to EPO agent(2 patient had bone marrow fibroses due to tertiary hyperparathyroidism and one had pure red cell aplasia probably due to EPO antibody) but 5 subjects (62.5%) had inadequate dosage of EPO.

**Conclusion:** EPO unresponsiveness is frequent and need full investigation including through history taking to find the underlying reason..

**Keywords:** Erythropoietin-stimulating agents , Resistant, Anemia, Etiology , Evaluation

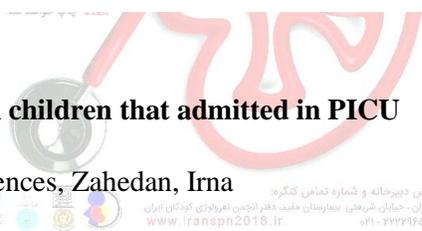
	Case1	2	3	4	5	6	7	8
age	10	17	10	1	14	1.5	8	2
Gender	male	male	female	female	female	male	male	male
WT/HT	51/120	56/157	40/120	8/60	32/150	9/60	52/125	9/80
Underlying disease	<u>vur</u>	<u>Vur</u>	<u>Nephrotic syn.</u>	Hus	Single kidney <u>vur</u>	<u>Hyperoxaluria</u>	<u>Nephrotic syn.</u>	<u>Nephrotic syn.</u>
Duration of dialysis	7y	10y	3y	3m	2y	2m	5y	1y
Type of dialysis	5periton 2 <u>hemo</u>	<u>hemo</u>	peritoneal	<u>hemo</u>	<u>Hemo</u>	peritoneal	<u>hemo</u>	<u>hemo</u>
EPO dosage/kg/ <u>wk</u>	4000 2weekly	10000 3weekly	2000 1weekly	2000 ½weekly	4000 weekly	2000 ½ weekly	2000 3weekly	2000 ½ weekly
BUN/Cr	60/9.8	50/9.5	70/10.2	50/7	80/9.8	50/8	60/10.2	50/8
HB	5.1	5	5.3	5.9	7.9	9.5	5.3	6
MCV	86	88	94	78	85	61	81	85
HCT	19	13	17.4	15	18	20	16.7	19.8
RBC	2.4	2.5	2.2	2.4	2.4	5.2	2.5	2.2
ACEI/ARB	-	-	-	-	<u>Enalapril</u>	-	<u>enalapril</u>	-
<u>Ca/P</u>	8.7/6.2	7.7/2.3	9.4/4.3	8/8.4	8.1/5.5	8.7/3.8	7.7/7.5	7.2/5.6
PTH	478	1857	456	289	270	1172	61	486
CRP	-	-	-	-	-	-	-	-

**P 14 :**

**Frequency of hypomagnesemia in children that admitted in PICU**

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**Background:** Magnesium is the fourth most abundant cation in the human body only after sodium, potassium and calcium and is the third intracellular cation. Magnesium is essential for human body health for the reason that ionized magnesium is involved in the interaction of more than 300 enzyme reactions and is important for electrolyte homeostasis, membrane stability, cell division, and generation of action potentials.

hypomagnesaemia is one of the most common electrolyte disturbances in hospitalized patients (12%), especially in the critically ill. The incidence of hypo-magnesemia varies from 20% to 65% in patients admitted to ICU.

**Methods:** In this study we searched about serom levels of magnesium in 150 children admitted to the PICU of Aliebne Abitaleb peace be upon him hospital and its relationship with outcome and risk factors. We found information with data forms and used SPSS software to analyse this data.

**Result:** In this study the incidence of hypomagnesemia in first day of hospitalization was 35.3% and incidence of hypomagnesemia in dead patients was more than others.

**Conclusion:** Hypomagnesemia in patients admitted to intensive care unit is common and have relationship to outcomes.

**Keywords:** Hypomagnesemia, PICU, outcome

## P 15:

### Occurrence of HUS in 13 months old infant infected with chicken pox

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Hemolytic uremic syndrome (HUS) is one of common cause of acute renal failure in children. HUS has variety of causes such as infections, genetic disorders, systemic diseases and drugs but occurrence of HUS after chicken pox is very rare.

Our case is a 13 months old boy that affected chicken pox. Diagnosis of chicken pox was clinically based on characteristic rash and distribution of lesions. One week after being infected, when skin rash and vesicles started to heal, he got into oliguria and edema. Further evaluation showed microangiopathic hemolytic anemia, thrombocytopenia and rise in BUN and Cr.

Patient with diagnosis of HUS got supportive care and renal function gradually improved during one week without any sequel in last 6 month follow up.

**Key word:**

Chicken pox - HUS – infant

## P 16:

### Investigating the Factors Affecting Tendency to Body Organ Donation: A Case Study at a Military University

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**Background and Aim:** Organ donation is a social action that is based on the individual's decision in time of life or Family consent at the time of brain death. The aim of this study was to investigating the Factors Affecting Tendency to Body Organ Donation: A Case Study at a Military University.

**Materials and Methods:** This descriptive cross-sectional study was conducted on 330 employees of a military university in 1396. The data were collected by questionnaire and face to face interview with two groups of cardholders without a donation card and analyzed using SPSS 18 software and descriptive and inferential statistics.

**Results:** According to Chi-square and t-test, there was a significant relationship between the organ donation and the two variables of age and education ( $P = 0.0001$ ). Results show that most of people (about 75 percent) has positive attitude to organ donation and persons with donation card had more share than person without card. Results of regression analysis show that with growth of rationalism, ethics, awareness of conditions and characteristics of organ donation and finally usage of communication media increase the probability or chance of that a person receives donation card.

**Conclusion:** The present study shows that social and cultural factors have a significant role in the donation and transplantation, and in order to pave the path of donation, a good look at these factors can be beneficial.

**Keywords:** social action, Rationalism, Ethics, Trust, Knowledge and awareness, universalism, Organ donation and transplantation

**P 17:****Central Nervous system Lesion shortly post renal transplantation- Difficult case**

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This was a -14 years old girl ,ESRD due to renal hypoplasia /dysplasia, received renal transplant from Unrelated donor (30 yrs old)/ no HLA study/D+/R+ for CMV-IgG,EBV IgG) since 2.8.2012.

The 1<sup>st</sup> wk post TX because of rising of creatinin from 1.5 mg/dl to 2.6 mg/dl she treated with MP and ATG with diagnosis of Acute rejection. Two weeks later she she presented with fever (T-38.5 C), vomiting, suicidal thought, aggressive behavior, depression and mood changes .CMV –Ag was positive (viral load  $470 \times 10^3$  copy)-Treated with: GCSF+ Meropenem + Vancomycin + IVIG+ gancyclovir+ valcyte + Floxetine but after two weeks of treatment gancyclovir stopped for bone marrow suppression and severe leucopenia. Afterward she admitted for another five times for abdominal pain, anorexia, mood changes. Medication was Cell cept (750+500)+ sandimun (75+100)+ Prednisolon (7.5)+COTMX. Four months post transplant she admitted again for Cough + rhinorhea , weight loss (3 kg), nausea, vomiting, anorexia, fever (38 C),staring and seizure (3 episodes) . CSF was nl, cultures were negative , brain MRI and brain CT scan – NL, CNS PCR was positive for EBV and CMV and blood CMV PCR load was 1920. Blood study showed EBV-Ab-VCA-IgG was positive ,IgM negative, and JC-BK virus positive She received IVIG + antibiotics + gancyclovir. Subsequently , she felt miserable , had mood disorder and depressed and sometimes lethargic.

The control Brain MRI one month later while she was still hospitalized showed progressive patchy area in hemisphere and basal ganaglia and cerebellum. The 2<sup>nd</sup> CSF PCR was negative for CMV, EBV, HSV, and Blood PCR was negative for toxoplasmosis DNA and Cryptococcus. AZT stopped and IVIG was given. But The neurologic symptoms and signs were progressive, she had high fever, needed mechanical ventilation, had GI hemorrhage, and had pleural effusion. Lung Ct scan showed nodules,

Third MRI of Brain showed the progression of the previous patchy lesions. Consult with hematologist recommended Rituxiamb IV and IT. The patients passed away five days later despite those treatment.

Conclusion- The possibility of Brain Lymphoma due to EBV was high. Unfortunately there were no tissue samples and the interpretation according serology and PCRs were problematic. Judies use of immunosuppressive is important to keep the EBV under control

**P 18:****Long term Outcome of Children on Hemodialysis –Single Center**

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**Aim:** To audit the performance of hemodialysis (HD) unit and to assess the outcome of children.

**Methods:** This is a retrospective cohort of children on HD in Ali-asghar children hospital between 2014 and 2017. Demographic data, clinical signs, HD treatment, and final outcome of children were reported. Data presented as percentage.

**Result:** During the past three years eleven patients went on chronic HD, Table below showed demographic data, access management, dialysis management. Three patients transferred to other center (adult unit –one case, and the other three transferred to pediatric unit near to their place of residence). One patient went on renal transplantation.

**Conclusion:** This is the first audit of HD of our center, We hope that serial assessment would help to improve the outcome.

Demographics	AACH
<b>Number of HD patients treated within the last 3 years</b>	8
<b>Age distribution</b>	
0 - 5.99 years	0 ( 0.0% )
6 - 12.99 years	4 ( 50.0% )
13 - 17.99 years	1 ( 12.5% )
> 18 years	3 ( 37.5% )
<b>eGFR at initiation of HD:Mean ± SD</b>	6.2 ± 1.7
<b>Comorbid conditions - Patients with:</b>	
Defined syndrome	1 ( 12.5% )
Impaired cognitive development	0 ( 0.0% )
Impaired motor development	2 ( 25.0% )
Ocular abnormalities	0 ( 0.0% )
Hearing abnormalities	1 ( 12.5% )
Cardiac abnormalities	1 ( 12.5% )
Pulmonary abnormalities	0 ( 0.0% )
Osseous abnormalities	3 ( 37.5% )
Other abnormalities	1 ( 12.5% )
<b>Access Management</b>	
<b>Distribution of access types (current)</b>	
Patients with central venous line (CVL)	6 ( 75.0% )
Patients with arteriovenous fistula (AVF)	2 ( 25.0% )
Patients with arteriovenous graft (AVG)	0 ( 0.0% )
<b>Mean number of hospitalization days related to HD access placement (Mean ± SD)</b>	3.1 ± 3.5 days
<b>Access related complications</b>	
AVF Dysfunction (1 event per X patient years)	29.2
<b>Dialysis Management</b>	
Number of dialysis sessions/week (Mean ± SD)	2.8 ± 0.5
Weekly dialysis duration (Mean ± SD)	9.8 ± 2.5 hours
Blood flow rate (Mean ± SD)	179 ± 61 ml/min/m <sup>2</sup> BSA
Kt/V urea per session (Mean ± SD)	2.35 ± 1.39
Kt/V urea per week (Mean ± SD)	6.66 ± 4.25
Anuric patients (<100 ml/m <sup>2</sup> /day)	3 ( 37.5% )
Daily urine output in non anuric patients (Mean ± SD)	1018 ± 330 ml/m <sup>2</sup> /day
Dialysate sodium (Mean ± SD)	140.5 ± 0.9 mmol/l
Pre-dialysis blood pressure systolic / diastolic (Mean ± SD)	127 ± 25 / 80 ± 19 mmHg
Number of antihypertensive drugs/patient (Mean ± SD)	2.9 ± 1.7
Dialysate buffer concentration (Mean ± SD)	33.5 ± 2.6 mmol/l
Venous HCO <sub>3</sub> (Mean ± SD)	19.6 ± 5.4 mmol/l
Nr. of complication episodes / 100 dialysis sessions (total)	1.59 (n=17)
- thereof	
- Hypotension	0.28 (n=3)
- Seizures	0.00 (n=0)
- Vomiting	0.09 (n=1)
- Muscle cramps	0.56 (n=6)
- Bleeding episodes	0.66 (n=7)
<b>Growth and Nutrition</b>	

BMI SDS (Mean ± SD)	0.3 ± 2.1
Malnutrition (BMI < 5th pct.)	1 ( 12.5% )
Overweight (BMI > 85th pct.) / Obesity (BMI > 95th pct.)	3 ( 37.5% ) / 1 ( 12.5% )
Patients with enteral feeding (NG tube + gastrostomy)	0 ( 0.0% )
Patients with intradialytic parenteral feeding	0 ( 0.0% )
Serum albumin (Mean ± SD)	37.7 ± 4.3 g/l
Height SDS (Mean ± SD)	-3.7 ± 0.6
Patients with height < 3rd pct.	7 ( 87.5% )
Patients receiving growth hormone	0 ( 0.0% )
Patients < 16 years with height < 3rd pct. receiving growth hormone	0 / 4 ( 0.0% )

**P 19:**

**What percentage of children on HD and CAPD met CKD-MBD KDOQI guideline?**

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**Background and Aim:** In children with end-stage kidney disease, one of the renal replacement methods including hemodialysis, peritoneal dialysis and kidney transplantation are selected for survival. In the dialysis process, the serum ca, Pi, and PTH levels are variable. Ca-ph levels increase in these patients, resulting in calcification, and calcium absorption of the gastrointestinal tract in CKD patients decreases. Chronic acidosis also reduces bone density by increasing calcium intake from the bone. KDOQi (2003) recommends that the ca-ph product be kept below 55 mg / dL. In this study, according to the standard National Kidney Foundation (KDOQi) a comparison of the adequacy of life of hemodialysis patients and peritoneal dialysis based on the changes in blood Ca-Ph and PTH was investigated.

**Method:** This was a retrospective cohort study of children under 18 years who were under treatment in Ali-Asghar children hospital between 2014 and 2017. During this period, the level of Serum calcium, phosphorus, and parathyroid hormone were measured. Data presented as mean (SD) and the percentage of patients who met the K/DOQI guideline.

**Result:** Eight patients were under hemodialysis and 56 were on CAPD during past three years. The detail of calcium (ca), phosphorous (Pi), PTH were shown in Table. Although mean calcium was similar in two groups but CAPD patients had higher Pi and PTH level.

**Conclusion:** This study shows that hemodialysis method is more effective in controlling the level of PTH in KDOQi PTH guidelines than peritoneal dialysis.

K/DOQI Target range	HD N=8	PD N=56
Serum Ca-mean(SD), mg/dl	8.82(1.2)	8.7(1.4)
Ca (%)		
Below		45.5%
Within		31.8%
Above		22.7%
Serum Pi -mean(SD),mg/dl	5.57 (1.85)	6.1(2.1) mg/dl
Pi (%)		
Below	28.6%	9.1%
Within	14.3%	54.5%
Above	57.1%	36.4%
PTH-mean(SD) , pg/ml	338(271)	646.3(589.1)
PTH (Target: 200-300pg/ml)		
Below	42.9%	31.8%

Within	14.3%	9.1%
Above	42.9%	59.1%
Patients on any phosphate binders	6 ( 75.0% )	100%
Patients on calcium-based phosphate binders only	2 ( 25.0% )	75%
Patients on calcium-free phosphate binders only	0 ( 0.0% )	41.7%
Patients on both calcium-based and calcium-free phosphate binders	4 ( 50.0% )	50%
Patients on oral calcium	1 ( 12.5% )	12.5%
Dialysate calcium (Mean $\pm$ SD)	1.3 $\pm$ 0.0 mmol/l	-
Patients on vitamin D3	1 ( 12.5% )	33.3%
Patients on active vitamin D analogue	6 ( 75.0% )	85.8%
Patients on cinacalcet	2 ( 25.0% )	6.25%

## P20:

### بررسی اختلال عملکرد توبولی در بیماران پیلونفریت تحت درمان با آمیکاسین

علی مهدوی - دانشگاه علوم پزشکی ایران ، تهران ؛ ایران

**مقدمه:** پیلونفریت یکی از بیماریهای شایع اطفال با شیوع ۷/۵-۴/۱ می باشد. بهترین درمان آنتی بیوتیکی برای پیلونفریت آمپی سیلین همراه با آمینوگلیکوزید وریدی است. آمینوگلیکوزیدها که سر دسته آن، جنتامایسین می باشد با اختلال عملکرد توبولی کلیه میزان دفع ادراری سدیم پتاسیم، کلسیم، فسفر و منیزیم را افزایش می دهد. بررسی اختلال عملکرد توبولی در بیماران پیلونفریت تحت درمان با آمیکاسین در سنین اطفال بین ۲ ماه تا ۱۲ سال انجام نشده است.

**روشها:** در این مطالعه کارآزمایی بالینی (Clinical Trial) تمامی بیماران مبتلا به پیلونفریت در گروه سنی از ۲ ماهی تا ۱۲ سالگی، بستری در بخش کلیه بیمارستان حضرت علی اصغر (ع) در طی شش ماه دوم سال ۱۳۸۳، در دو گروه جداگانه به صورت راندوم تحت درمان با آمپی سیلین و جنتامایسین، آمپی سیلین و آمیکاسین مورد بررسی قرار گرفته اند. نمونه راندوم ادرار و سرم قبل از شروع درمان روز دوم و پنجم و هشتم درمان از بیماران گرفته و الکترولیتها، Cr و اسمولالیتیه ادرار و سرم اندازه گیری شد.

**نتایج:** با بررسی سیر مقادیر، بین روزهای ۰ و ۲ و ۵ و ۸ بعد از درمان میانگین سطح سرمی و اندکسهای ادراری الکترولیتها تفاوت آماری معنی داری در هر گروه و در مقایسه با هم وجود نداشته است، به جز میانگین کسر دفعی کلسیم FE Ca که بین این روزها اختلاف آماری معنی داری داشته است. ( $\text{sig} <$  Within subjects 0.05) هر چند تغییرات این میانگین در طی زمان بین این دو گروه اختلاف معنی داری نداشته است.

بحث: در مطالعه ما نیز مشابه مطالعات دیگر بیشترین افزایش در دفع کلسیم در ادرار در طی ۲ روز اول درمان (به صورت اثر زودرس بعد از تجویز آمینوگلیکوزیدها) با هر دو آنتی بیوتیک بوده است.

## P21:

### استفاده از واکسیناسیون در پیشگیری از عفونت ادراری

دکتر رضا دلیرانی

فوق تخصص نفرولوژی کودکان، بیمارستان کودکان مفید، دانشکده علوم پزشکی شهید بهشتی

از روش های معمول و شناخته شده در پیشگیری از عفونت ادراری استفاده از آنتی بیوتیک پروفیلاکسی میباشد. با توجه به عوارض دارویی و احتمال مقاومت آنتی بیوتیکی و عود عفونت لزوم پیدا کردن روشهای جایگزین در پیشگیری از عفونت ادراری اهمیت بسزایی دارد. این روش ها ی پیشگیری شامل استفاده از پروبیوتیک ها، اصلاح اختلال دفعی ادرار و مدفوع، انجام ختنه و اعمال جراحی رفلکس وزیکوپورترال ، استفاده از cranberry و موادی که مانع چسبیدن باکتری به سطوح اوروپیتلیوم میشود، اخیرا استفاده از واکسن برای پیشگیری از عفونت ادراری مورد استفاده قرار گرفته است.

E-coli یوروپاتوژنیک (UPEC) شایعترین ارگانسیم ایجاد کننده عفونت ادراری میباشد. در پاتوژنز UPEC نقش گیرنده های متعدد مورد بررسی قرار گرفته است که در بین آنها دو نوع گیرنده type I و Ppili اهمیت بیشتری دارد. فیمبریای type I در پاتوژنز سیستمیت و نوع P در ایجاد پیلونفریت نقش دارند. تداخل در اثرات این گیرنده بر سلول های اوروپیتلیال حیطه مورد استفاده در درمان داروها و واکسن های جدید میباشد.

تاریخچه استفاده از واکسن در عفونت ادراری بی بیش از یک قرن میرسد. اولین استفاده در سال ۱۹۰۹ بیشتر به عنوان استفاده درمانی تا پیشگیری بود. برای دهه های بعد نتایج استفاده از واکسن به منظور درمان موفقیت آمیز نبود. از سال ۱۹۵۰ استفاده از واکسن به عنوان پیشگیری در عفونت ادراری مورد توجه قرار گرفت. تاثیر واکسن در مدل های حیوانی در انواع واکسن (کنژوگه ، توکسویید، غیر فعال شده و ضعیف شده) نشان داده شده است. از واکسن های جدید که در مدل های انسانی آزمایش شده ، فقط یک نوع آن به صورت تجاری در دسترس میباشد.

واکسن های عفونت ادراری در دو گروه کلی قرار دارند. ۱- واکسن شامل کل ارگانسیم (whole cell) که به صورت زنده ضعیف شده یا غیر فعال است. ۲- specific antigen شامل یک یا چند آنتی ژن (subunit، توکسویید یا واکسن های کنژوگه). واکسن های specific antigen عموما از واکسن های کامل بی خطر تر هستند. واکسن موثر باید سطوحی که به وسیله یوروپاتوژن در مراحل مختلف عفونت مورد حمله قرار میگیرد هدف قرار دهد و از اعمال اثر پاتوژن جلوگیری کند. استفاده از واکسن ها در دو گروه پیشگیری از سیستمیت و پیلونفریت مورد توجه قرار گرفته است.

تنها واکسن عفونت ادراری که در حال حاضر برای انسان استفاده میشود واکسن کل ارگانسیم (whole cell) غیر فعال شده پلی والانته به نام Strovac میباشد. این واکسن شامل ۱۰ گونه مختلف است. در سال های گذشته واکسن دیگری به نام Solco urovac در اروپا استفاده شد که آن هم شامل ده گونه (۶ تا گونه E-coli و یکی کلبسیلا، پروتئوس، مورگانلا و انتروکوک) بود.

**P 22:****Impacts of chronic functional constipation on bladder ultrasonography: a case- control study**

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**Introduction:** Association of chronic constipation with vesicoureteral reflux, urinary tract infection, and dilatation of upper system have been reported. We aimed to define the impact of chronic constipation on bladder sonography indexes (bladder volume, full and empty bladder wall thicknesses, and urinary residual volumes after once and twice bladder emptying).

**Methods:** bladder indexes were compared between cases with chronic constipation aged  $\geq 5$  years and control group (those with normal urinary and defecation patterns). Urinary residual volumes after once and twice bladder emptying  $>30$  and  $>20$  ml in children  $\leq 7$  years and  $>20$  and  $>10$  ml in children  $> 7$  years were considered abnormal respectively.

**Results:** 32 children with chronic constipation and 69 normal cases enrolled the study. Comparison of full bladder volume (cc), full and empty bladder wall thicknesses (mm) and post void residual volumes after once and twice bladder emptying didn't show significant differences ( $P=0.53, 0.215, 0.346, 0.502$  and  $0.352$  respectively), although full and empty bladder wall thicknesses and post void residual volumes after once and twice bladder emptying were higher in cases with chronic constipation.

**Conclusion:** chronic constipation has no significant impact on bladder volume, post void residual volume and bladder wall thicknesses indexes in full and empty condition. Whether functional bladder capacity affects by chronic constipation, uroflowmetry test and recording frequency volume chart in cases with chronic constipation and comparing them with normal children may add our knowledge.

**Key words:** Chronic functional constipation, bladder US, Bladder volume, bladder wall thickness, post void residual volume, children

**P 23:****Compression between Normal Saline & hypertonic Saline in inducing hyponatremia after surgery**

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**Introduction:** Hyponatremia is the most common electrolyte disorder in patients following surgical interventions (19-50%). Hospital acquired hyponatremia is often due to using hypotonic solutions and can be lethal.

**Materials and Methods:** Between January and December 2014, 190 children (1 month to 12 years) who were admitted in the urology department of Children's Hospital Medical Center for elective surgical procedures were enrolled in the study. The patients were randomly divided into two groups: group I received 50 mEq/L sodium and 20 mEq/L potassium in D/W 5% and group II received 154 mEq/l sodium and 20 mEq/L potassium in D/W 5% at the maintenance dose for a period of 6 hours following the operation. The patients did not have any oral fluid intake 6 hours postoperatively. The incidence of hyponatremia before and after maintenance IV fluid therapy was analyzed. Other characteristics of the patients such as age, gender, duration of hospitalization, other concomitant electrolyte disturbances, and symptoms of hypervolemia were also evaluated. The incidence of fluid-IV therapy-induced hyponatremia

was investigated and analyzed in different categories of patients.

**Results:** One hundred and ninety patients were enrolled. The mean age was 3.75 years (ranging from 1 month to 12 years). One hundred and thirty-three patients (70%) were boys. The incidence of hyponatremia before and after maintenance IV fluid therapy was 9.5% and 36%, respectively. After the therapy, the incidence of hyponatremia was 54% and 17% in hypotonic and isotonic groups, respectively. Final multivariate logistic analysis showed that hyponatremia was common in patients that received hypotonic solution after surgery.

**Conclusions:** Hyponatremia was markedly induced in patients receiving hypotonic solution after surgery. It seems isotonic fluid therapy after surgery protects the patients from hyponatremia.

Keywords: Hyponatremia; Isotonic solutions; Hypotonic solutions; Child.

## P 24:

### The evaluation of the effect of the length of Induction course of the childhood idiopathic nephrotic syndrome on relapse risk

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Idiopathic nephrotic syndrome (INS) is characterized by relapsing proteinuria with potentially 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 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.

**Method and materials:** All new cases of childhood INS selected. Oral prednisolone at 2mg/kg/d(max.60mg), as 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 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: induction phase  $\leq 1$  week (9 patients), G2:  $>1$  week  $\leq 2$  weeks (8 patients) G3:  $>2$  weeks  $\leq 3$  weeks (8 patients) and followed 2 years for relapse rate. Patients

**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 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. ( $\rho$ .value=0.43, =0.71&=0.73, respectively).

**Conclusion:** 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.

Keywords: nephrotic syndrome, induction, relapse, children

**P 25:****Bone mineral density in children with relapsing Nephrotic Syndrome**

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**Introduction:** Given the high relapse rate of disease in children with steroid dependent nephrotic syndrome and the osteoporotic effect of long periods of steroid therapy, this survey was performed to find the bone mineral status of these patients.

**Methods:** Bone mineral density and content (BMD and BMC) were measured using Dual energy X-ray absorptiometry in 37 nephrotic children, six girls and 31 boys aged from four to 21- yrs, as patient group and 37 age and sex-matched healthy individuals as control group. Historical data were collected by chart review.

**Results:** As compared to the control group, the patients were shorter in stature. The percentage of BMC of lumbar and BMD of femoral bones of the patients was significantly lower than control group. According to the Warner method, 12% of the patients were osteoporotic and the BMD of their femoral and lumbar bones was inversely correlated with cumulative steroid dose.

**Conclusion:** Bone loss can occur in some steroid-dependent nephrotic patients, especially those with low age of onset and those with longer duration of the disease and higher cumulative dose of steroid. Therefore, measurements of BMD and BMC could be recommended, at least, for the selected patients

**P 26:****Status epilepticus :a clinical presentation of hypertensive encephalopathy**

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**Back ground:** Status epilepticus is a serious neurologic emergency during childhood period. Major causes include hypoglycemia, head trauma, intracranial infections, hypo and hypernatremia, hypocalcemia, hypomagnesemia, drug withdrawal and intoxications. Some encephalopathies due to uremia, hypertension and dialysis may lead to seizures. This study presented two cases with hypertensive encephalopathy and status epilepticus.

**Methods:** Two cases with status epilepticus were admitted in Ali-Asghar children hospital during 2016-2017. Data about age, gender and clinical manifestations was recorded.

**Results:** First case was a 5 year old girl who was admitted in our intensive care unit with status epilepticus. Seizures was refractory to antiepileptic therapy ,Phenobarbital ,phenytoin and miadazolam. When hypertension was detected, after antihypertensive treatment, seizures completely controlled. During hospitalization, we found that she regressed in motor, speech and cognition and had no visual attention. Electroencephalography detected rare paroxysmal discharges. There was not significant abnormality in brain CT scan. Renal biopsy showed focal and segmental glomerulosclerosis and treatment was started. Gradually motor-speech delay and visual function became near normal. Second case was an eight year old boy with autoimmune encephalopathy and received high doses of intravenous methylprednisolone for five days. One week after recovery, he experienced headache, restlessness and generalized seizures that continued and was finally status epilepticus. Hypertension and abnormal signals in occipital region of the brain-in magnetic resonance imaging-was suggested posterior reversible

encephalopathy. After treating of hypertension that was supposed to corticosteroid medication, clinical findings subsided.

**Conclusion:** Hypertensive encephalopathy may cause loss of consciousness, cognition impairment and seizure disorder. In our cases hypertension was presented with status epilepticus.

### P 27:

#### **Pulmonary Embolism in pediatric nephrotic syndrome**

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**Introduction and aim:** Pulmonary Embolism (PE) is a known complication of Nephrotic syndrome (NS) specially in relapse period.

Frequently PE is asymptomatic and the diagnosis is not simple and needs to be suspicious to. Incidence of PE in children with NS is recognized less than adult patients and under recognized with a high rate of mortality and pulmonary hypertension.

Chest CT Scan and MRA are the best diagnostic exam but lung ventilation perfusion Scan is the most available diagnostic test. Due to the H. Dirk Sotsman and colleagues, chest X Ray can be used instead of ventilation scan.

In this study we are going to find asymptomatic pulmonary embolism in pediatric nephrotic syndrome.

**Method:** For Six children 2.5-8.5 years of age suffering of NS and hospitalized because of NS relapse, perfusion scan and CXR were done. They did not show any sign or symptom for PE. Perfusion scan was read in comparison to CXR and the result reported as high, moderate and low probability for pulmonary embolism.

**Results:** 5 patients had normal CXR and perfusion scan, and one patient had a positive perfusion scan and a normal CXR. The perfusion scan demonstrated no perfusion in the left upper lobe. She was a 4.5 years old girl with serum albumin about 2mg/dl and serum triglyceride about 185 mg/dl. Treatment with Heparin was started. The Control Perfusion Scan after one month was normal.

**Conclusion:** PE as a complication of NS is underestimated and there are some subtle cases that cannot diagnose them without performing special diagnostic tests. For prevention of the PE mortality and morbidity in pediatric NS, it needs to be vigilance.

**Key words:** nephrotic syndrome, Pulmonary embolism, perfusion scan.

### P 28:

#### **Serum interleukin 13 level in steroid sensitive NS**

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**Introduction:** There are studies indicating the relationship between interleukin 13 and pathogenesis of nephrotic syndrome. To find this relationship, we measured serum IL13 concentration in acute and remission phases in children with steroid sensitive nephrotic syndrome.

**Methods:** The serum of 15 patients was achieved for measurement of IL-13 in two phases: acute phase and remission phase. To this end, we used Biofarm ELIZA kites.

**Results:** The mean concentration of serum IL-13 was 430\_279 ng/mL (227 - 960 ng/mL) before starting steroid treatment. The mean concentration of serum IL-13 was 428\_259 ng/mL (215.6 - 960 ng/mL) after 4 weeks of steroid treatment. The difference between pre and post treatment of serum IL-13 levels was not significant ( $P = 0.91$ ).

**Conclusions:** We showed serum IL13 does not significantly decrease after steroid treatment. Although the role of IL13 in increasing glomerular permeability has been shown in previous studies, the local IL13

may be more important than the systemic form. It also can be postulated that mediators other than IL13 also have primary roles in pathogenesis of proteinuria. However, we need more studies to determine the role of IL13 in the pathogenesis of nephrotic syndrome

### P 29:

#### Compare serum levels of interleukin-6 (IL-6) in acute pyelonephritis versus acute cystitis in 6 months to 12 years old children

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**Introduction :** UTI is a common disorder in pediatrics and early diagnosis is important . However in young children diagnosis facing a problem which makes diagnosis hard .

One of them is variety in sign and symptoms and for exact diagnosis in some case need expensive modalities . So new way is needed . il-6 at last decades preferred to

UtI diagnosis and upper / lower involvement differentiation. So according to contrariety in IL-6 accuracy of plasma IL-6 in diagnosis and its power for differentiating upper from lower involvement.

**Material and methods:** at this descriptive study , 83 patients were included and examined for interlukin - 6 value in both urinary tract diagnosis and upper/lower involvement differentiation . Demographic characteristics like age , gender , history , physical examination beside of blood work like white blood cell count , ESR, CRP , IL-6 done and listed . Descriptive parameters report by mean and frequency for statistical analysis done t- test in SPSS16 . ROC curve downs for diagnosed value .

**Results :** in this study 83 patients ( 14 control group , 33 ( 39.8%) cystitis , 36 ( 43.4%) pyelonephritis with 3.93+\_3.069 mean age enrolled .

Age distribution in this study 20 , 63 for male and female respectively . The interlukin -6 level in cystitis was higher than control and but this is not significant . In ROC curve sensivity and specificity of IL- 6 for cystitis 52% , 50% and for pyelonephritis 60% , 50% respectively .

**Conclusion :** according to this study , IL-6 isnt a good Biomarker for UTI and differentiation Upper Tract involvement from the Lower Tract , A wide range of IL-6 level and useless of this Biomarker can be due to gender distribution ,time of diseasepresence , bacterial virulence and other factors . So suggestion for future studies on the large sample which parameters like gender distribution and other one mentioned controlled tightly.

### P 32:

#### Nephrocalcinosis in preterm neonates, Afzalipourhospital, Kerman university

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**Introduction:** Nephrocalcinosis is defined as renal calcification. Low gestational age and low birth weight, male sex, some drugs such as furosemide, dexamethasone, methyl xanthines, steroids, excess vitamin D and calcium intake by mother are some of its known precipitating factors. Some studies suggested mechanical ventilation, the duration of hospital stay and TPN as precipitating factors.

The aim of this study was to assess the frequency of some of these precipitating factors regarding nephrocalcinosis in premature neonates.

**Materials and methods:** This cross-sectional study was conducted following an approval from the ethics committee of Kerman university of medical sciences in which 115 premature infants less than 32 weeks of gestation admitted in NICU Afzalipour hospital, Kerman, Iran were assessed during years 2012-2013. Data regarding neonates including sex, age, weight, apgar score at the time of birth, hospitalization duration, prescribed drugs, with or without mechanical ventilation and lab data were gathered by

checklist. Renal ultrasound was obtained from all patients at the end of 8<sup>th</sup> week of age by a professional radiologist and by the same device. All data were entered and analyzed by descriptive tests using spssv.17 and the results were reported as mean±SD for quantitative data and frequency for qualitative data.

**Results:** mean gestational age at the time of admission was 30-35 weeks. 50 neonates were female and 65 were male. Nephrocalcinosis was seen in 42 (36.5%) of the patients. Nephrocalcinosis was more common in males and was associated with family history of renal stone, high urinary calcium, low urinary citrate, plasma high calcium levels, vancomycin prescription in neonate and intake of excess vitamin D and calcium by mother respectively. In contrast, no association between nephrocalcinosis and gestational age, apgar score at the time of birth and usage of aminoglycoside, dexamethasone and mechanical ventilation was observed.

**Conclusion:** demographic indices and nephrocalcinosis precipitating factors assessed in the present study was in accordance with other available studies so far, but our results regarding vancomycin prescription and family history of renal stones in association with nephrocalcinosis were in contrast to other studies.

Keywords: nephrocalcinosis, hypercalciuria, premature neonate, vitamin D

### P 33:

#### Comparison of serum and urinary cystatin-c in in children with type one diabetes mellitus and controls

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Aim of study: cystatin- c is recently introduced for estimation of GFR in diabetic patients. Metabolism and factors impacting serum and urinary levels of cystatin- c is less studied, especially in children. Here we compared serum and urinary level of cystatin-c in in children with type one diabetes mellitus and controls to answer the question that is cystatin-c production and elimination is altered in diabetes mellitus?

Method: patients with stable T1D and healthy controls aged 2–18 years with no history of recent infection were included. Written informed Consents were taken from the parents before enrollment. Cystatin-c were determined in fasting plasma sample and 24 hours urine. Other variables were age, gender, weight and height percentile, body mass index, thyroid status, blood pressure, HbA1C, serum creatinine, 24 hours urine albumin and GFR. Comparisons were performed using appropriate statistical approach

Results: plasma cystatin-c had normal distribution. Mean and standard deviation of plasma cystatin-c in diabetic and normal subjects were 896(358) and 937(183) microgram in liter, respectively. There was no significant difference among plasma level of cystatin-c between diabetic and normal subjects or diabetic with and without albuminuria. Urine cystatin-c was not normally distributed. Median and range of urinary cystatin-c were 22(999) for diabetic patients without proteinuria, 30(2492) for diabetic patients with proteinuria and 26(297) for normal subjects. Urinary creatinine had no significant difference among tree groups. Urinary volume, as expected, was higher in diabetic patients, both without and with albuminuria, than normal ones. Diabetic patients had higher plasma creatinine than normal subjects. GFR had no significant among tree groups.

Conclusion: despite similar plasma levels of cystatin-c, diabetic patients with albuminuria have more variations in urinary elimination of cystatin-c. cystatin-c seems to differentiate the renal failure before decrease of GFR.

Key words: cystatin-c, children, type one diabetes mellitus

## P34:

بررسی سطح سیستاتین - سی در پلاسما و عوامل تاثیر گذار بر آن در کودکان مبتلا به دیابت نوع یک

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- هدف مطالعه : مطالعه ی حاضر به منظور تعیین سطح پلاسمایی سیستاتین - سی در کودکان ایرانی مبتلا به دیابت نوع یک و عوامل موثر بر آن انجام شده است.
  - روش مطالعه : بیماران مورد مطالعه کودکان ۲ تا ۱۸ ساله مبتلا به دیابت نوع یک بودند. متغیر های مورد بررسی در این مطالعه شامل سن، جنس، قد، وزن، شاخص توده ی بدنی، فشار خون سیستولی و دیاستولی، طول مدت دیابت، مرحله ی بلوغ و وضعیت تیروئید<sup>۱</sup>، سطح سیستاتین سی، کراتینین سرم و هموگلوبین A1C در خون ناشتا، همچنین میزان آلومین، سیستاتین سی و کراتینین در ادرار بیست و چهار ساعته و میزان فیلتراسیون گلومرولی بود. ارتباط متغیر های مورد بررسی با سطح سیستاتین توسط آزمون رگرسیون چند متغیره ی خطی بررسی گردید.
  - نتایج مطالعه : در مجموع ۹۱ بیمار دیابت نوع یک وارد مطالعه شدند. میانگین سیستاتین سی در سرم  $896 \pm 358$  میکروگرم در لیتر بود. در آنالیز رگرسیون خطی چند متغیره مشخص شد عواملی که به طور معناداری بر سطح سرمی سیستاتین سی تاثیر گذارند وجود فشارخون دیاستولی و وضعیت تیروئید است و سایر موارد بررسی شده تاثیر معناداری ندارند.
  - نتیجه گیری : نتایج مطالعه ی حاضر نشان میدهد از بین عوامل مورد بررسی مهمترین عامل موثر بر سطح سیستاتین سی سرم، فشار خون دیاستولی و نیز وضعیت هورمون های تیروئیدی است. توجه به وضعیت هورمون های تیروئید و فشار خون در کودکان با دیابت نوع یک میتواند از تخمین نادرست وضعیت کلیوی با استفاده از سیستاتین-سی جلوگیری کند.
  - کلمات کلیدی : سیستاتین - سی، کودکان، دیابت نوع یک

## P35:

### Measurement Of Cystine In Granulocytes Using Liquid Chromatography-Tandem Mass Spectrometry In Iranian Children

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**Background and aim:** Cystinosis is a rare autosomal recessive disorder characterized by an accumulation of intra lysosomal cystine due to a defect in cystine transport across the lysosomal membrane. This results in cystine accumulation and crystallization in the cells causing widespread tissue damage. This disorder can be treated specifically using high doses of cysteamine. Although the pathogenesis is not completely elucidated, research in the field of cystinosis has been spurred in the late 1990s by the identification of the CTNS gene, which encodes for cystinosin, a lysosomal cystine carrier. Accurate measurement of intracellular cystine content is necessary for the diagnosis and monitoring of treatment with cysteamine. Here we describe a new method to measure intracellular cystine.

It relies on a liquid chromatography-tandem mass spectrometry assay. In the 1960s, Schneider et al. showed for the first time increased intracellular cystine levels in granular fractions of leukocytes from patients, identifying cystinosis as a cystine storage disease that was associated with Fanconi syndrome. Cysteamine (Cystagon) depletes lysosomal cystine by a disulfide exchange reaction, resulting in the equimolar generation of a cysteine-cysteamine molecule and a molecule of cystine. Both compounds can exit lysosomes via "system c" transporters, bypassing the defective cystinosin pathway. The efficacy of cysteamine can be monitored in clinical practice by measuring intracellular cystine levels in polymorphonuclear (PMN) leukocytes and is considered to be a reflection of tissue cystine content. Over the years, several methods have been developed to measure intracellular cystine levels for the diagnosis of cystinosis and for monitoring cysteamine treatment. Currently, HPLC and liquid chromatography followed by tandem mass spectrometry (LC-MS/MS) are the most widely used. LC-MS/MS is the most sensitive method (allows detection of as little as 0.02  $\mu\text{mol}$  cystine/l, compared with 0.15  $\mu\text{mol}$  cystine/l using HPLC) and has recently been developed for cystine determination in granulocytes.

**Materials And Method:** In order to measure final concentration of cystine in nanomoles of half cystine by milligrams of proteins it is necessary to measure the amount of cystine in lysed RBCs and the protein content of cell debris both analysis are equally important. 7-10 ml venous blood in ACD or heparin or EDTA or monitoring treatment sample should be collected 6 hr after cysteamine treatment was started. Whole blood is conserved at room temperature until sample is prepared. PMN cells are prepared within 24 hr according to guidelines from group "cystine in white blood cells". Isolation of PMNs increases the sensitivity for detection of heterozygotes after simple preparation and leukocyte isolation. Then 50  $\mu\text{l}$  of sample is injected into LC-MS/chromatographic separation is achieved on an Agilent 1100 system using a C18 ODB column. The column elute is injected directly into Applied Bio system API 13000 MS/MS. This method allows to quantify amount of cystine in lower than 0.25 nmol  $\frac{1}{2}$  cystine/mg protein in normal subjects. It is possible to measure very low concentrations of intracellular cystine with liquid chromatography-tandem mass spectrometry. The results obtained with this novel method correlate very well with those obtained using the cystine-binding protein assay.

**Results:** We analysed leukocyte cysteine level of 308 patients (146 Females and 162 Males) with mean age 8.5 yr (range 0.01 – 71 yr) referred to our laboratory between 2015-2017. The results were shown in the table. The mean cystin level of leukocytes was 0.64 (range 0- 6.54). Which shows a frequency of about 50% for patients with cysteine level  $>0.4$

### Frequencies

**cystin0.4**

	Frequency	Percent	Valid Percent	Cumulative Percent
Valid >0.4	155	50.3	50.3	50.3
<0.4	153	49.7	49.7	100.0
Total	308	100.0	100.0	

**Conclusion:** Since efficacy of cysteamine can be monitored in clinical practice by measuring intracellular cystine levels in polymorphonuclear (PMN) leukocytes and is considered to be a reflection of tissue cystine content we recommend this method for diagnosing of the disease and follow up of patients receiving Cystamine to evaluate the efficacy of treatment

### P36:

#### The performance of Plasmapheresis Unit in Ali-asghar children hospital- an audit

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**Background and aim:** Therapeutic plasmapheresis is a method in which a large amount of plasma is removed from the body and replaced by 4.5% plasma or albumin solution. It helps to reduce the concentration of proteins, fats, toxins, antigens and antibodies and immune complexes such as s lupus, which are involved in antibodies and immune complexes, it is possible to prevent kidney failure by removing antibodies. This is an audit of plasmapheresis unit in a children hospital.

**Methods:** This was a retrospective descriptive study conducted between 2016 and 2017 in Ali-asghar children hospital. All patients went under plasmapheresis with Optia or Feresenius devices. The replacement fluid was Fresh Frozen Plasma with the volume of 30-40 ml/kg for at least 3 to 5 sessions in hemolytic uremic syndrome or thrombotic thrombocytopenia. For other patients replacement fluid was a mixture of Albumin, FFP and Normal saline. Blood pump , citrate infusion ,and priming of catheter was per protocol.

**Results:** The number of patients (13 M, 13 F) who were treated with plasmapheresis was 26 with age of 9 months to 14 years. The range of weight was 9-50 kg. The duration of plasmapheresis in each session was 50-140 minutes. The majority of patients suffered of hemolytic uremic syndrome. The observed complications were hypocalcemia, Coldness and shiver , erythema , rash , Leakage of blood around exit site that happened in 9 children. Two sessions membrane were ruptured or device was out of order. Catheter malfunction, ineffective plasmapheresis were other complications.

**Conclusion:** this was the first report of plasmapheresis unit in Iran. The rate of complication was low and it was safe in small children

### P37:

## Pericardial effusion in End Stage Renal Disease (ESRD): Report of 5 cases and review of literature

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**BACKGROUND:** Pericardial effusion, common at one time among dialysis patients, has become a rare entity in recent years. Due to its low incidence, its recognition has gained importance among internists, cardiologists, and nephrologists. It can be seen in predialysis patients and in dialysis patients who are on hemodialysis or peritoneal dialysis.

**METHOD:** A retrospective study is performed reviewing all hospital admissions of the patients suffering from ESRD and pericardial effusion. The clinical manifestations, laboratory data, echocardiography and treatment modalities of the patients during each admission is recorded.

**CASE REPORT:** We report 5 cases of uremic pericarditis and their presenting manifestations and review the literature. Among these patients, the traditional findings of acute pericarditis such as chest pain, fever, electrocardiographic changes, and leukocytosis are uncommon. Muffled cardiac sounds have a relatively high incidence and should be emphasized on auscultation of uremic patients. Not infrequently, it is complicated by pre-tamponade or tamponade, requiring pericardiocentesis or pericardial surgery.

**CONCLUSION:** Uremic pericardial effusion is treatable, but not always a preventable, condition. Timely recognition of its presence and its efficient management are essential elements of successful treatment.

**Key words:** ESRD, pericardial effusion, clinical manifestations, treatment

P38:

## EBV infection after Renal Transplantation in Children- A report of Iran

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**Definition:** Epstein-Barr virus (EBV; now also known as HHV-4) is another DNA herpes virus. The patterns of infection are identical to CMV: primary infection (often from the graft of a seropositive donor), reactivation, or superinfection. Also, as with CMV, the primary infection in an immunosuppressed transplant recipient is more virulent.

**Epidemiology :** The rate of EBV infection in EBV seronegative pediatric transplant recipients (as defined by systemic viremia) ranges from 50% to 80%. The KDIGO guidelines recommend the following post transplant EBV schedule for high-risk D+/R-patient: once in the first week after transplant, at least

monthly for the first 3-6 months, then at least every 3 months until the end of the first year with re-initiation of monitoring after treatment for acute rejection.

**Diagnose:** Experience has shown that serology is unreliable as a diagnostic tool for either PTLD or primary EBV infection in immunocompromised patients. These patients show a marked delay in their humoral response to EBV antigens, and many fail to develop immunoglobulin (Ig) M antibodies altogether. Another important drawback is that these patients often receive blood or blood products with the passive transfer of donor antibodies.

In the case of EBV, testing can be performed against EB nuclear antigen or viral capsid. The former turns positive early, after recent infection and the latter in more distant infection. More recently, in western countries, PCR amplification of viral DNA is the method most commonly used.

**Treatment** There is no universally accepted treatment for EBV infection post transplant; Reduction in immunosuppression is one modality. The antiviral agents ganciclovir and valganciclovir have activity against EBV. Acyclovir is not effective against EBV. The duration and route of antiviral treatment are not standardized but most centers will treat until the EBV PCR has turned negative. Oral ganciclovir or valganciclovir are also used for prevention of EBV disease. These agents seem to delay the onset of infection rather than reduce the incidence.

**PTLD:** Unlike CMV, EBV infection does not seem to have many indirect effects except for the development of PTLD, which is an uncontrolled proliferation of immune cells (typically B cells) in the setting of posttransplant immunosuppression. In pediatric cases especially, most PTLD is due to Epstein Barr virus. This condition is not a true malignancy since the immune system can regain control of B-cell proliferation if extrinsic immunosuppression is reduced and EBV- directed CD8+ T cells regain function.

Posttransplant Lymphoproliferative Disorder (PTLD), 4-5 x more common in children after kidney transplant than adults. Usually caused by proliferation of Epstein Barr virus (EBV) infected B cells

Symptoms :Infectious mononucleosis ,Lymphoid hyperplasia,Invasive malignant lymphoma

Malignancy associated with polyclonal expansion of B cells associated with rise in EBV titers

Incidence of PTLD in pediatric renal transplants is 1.2% overall,Incidence has increased slightly

Increased incidence with use of tacrolimus, white race, and cadaver donor. Treatment generally involves reduction in immunosuppression dose and antiviral agents

PTLD can present clinically in many ways. Most often, the patient has lymphadenopathy, fever, or symptoms related to pressure on internal organs from an internal lymphoid mass.

Diagnosis is made histologically, though imaging can help with appropriate tissue acquisition and staging. Monoclonal or monomorphic PTLD is generally more severe than polyclonal or polymorphic. The most severe forms are indistinguishable from lymphomas. PTLD incidence in pediatric kidney transplantation was previously low at < 1% but then climbed over the last 10 years to more than 4%.

Treatment of PTLD usually involves a sequential process. Immunosuppression reduction is typically the first intervention, followed by the anti- CD20 antibody rituximab or interferon alfa.

Chemotherapy is usually reserved for higher – grade cases. Rituximab can be added to either full- dose or attenuated- dose chemotherapy with good results with either type of combination therapy.

**Method:** This is a cross sectional study using descriptive methods. in this research 20 Childs underwent transplantation and were investigated in term of age, gender, the cause of renal failure, EBV serology before and viral load after transplantation ,drug using before and after transplantation

**Result :** In total, out of 20 children 75% were boys and 25% were girls. The average age was 11.26. 70% of patients were D<sup>+</sup>R<sup>+</sup> and 30% were D<sup>+</sup>R<sup>-</sup>. Only 2 of them after transplantation had EBV viral load 400, 1300. Renal failures in these individuals were: poly cystic kidney disease, hypo plastic ,PUV ,Neurogenic bladder and FSGS. Most of these children didn't have a history of underlying disease. But among those who had, 1 person had ADHD, 1 had hypothyroidism and 4 of them had hypertension. Donor of renal transplantation had been often unknown, but from 11 persons that it was known 4 of them had cadaver donor. The average time passed from their transplntation wa 34.9 months .Immune suppressive therapy regimen was prednisolon,prograf,cellcept . there was no acceleration between gender and viral load of EBV. And also there was no acceleration between viral load of EBV and cause of renal failure, underlying disease.drug using and cadaver or living donor. Only there was acceleration between viral load of EBV and the duration time of transplantation.

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