Value of electron microscopy in establishing the diagnosis of early stage of renal amyloidosis

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Introduction: Amyloid represents the tissue deposition of fibrils derived from amino-terminal region of light and heavy chains of homogenous immunoglobulin. Most patients with renal amyloidosis have proteinuria. The kidney is frequently affected in different types of amyloidosis therefore the kidney biopsy is a diagnostic method. Renal amyloidosis is the major cause of death so diagnosis at early stages is very important to search for any cause of amyloidosis and possible early treatment of primary disease. The purpose of this study was to determine the number of patients with normal light microscopy and negative Congo red staining but positive amyloid fibrils by electron microscopy (EM) in the kidney biopsies.

Methods: A total of 32 cases of renal amyloidosis were diagnosed in our center by electron microscopy (LEO 906) in pathology department during 15 years. The specimen for EM were fixed in Glutaraldehyde 3% and after routine processing and preparing Resin blocks ultra-thin sections (90nm) were stained by Lead citrate and Uranium acetate and studied and photographed by EM.

Result: Electron microscopy showed masses of fibrilar material in the mesangial area of glomeruli with narrowing of capillary lumen. The fibrils were non-branching with diameter of about 8-12 nm. In some patients with advanced disease the fibril deposition were also observed around the blood vessels or basement membrane of tubules. In 5 (15.6%) patients the Congored stain for amyloid were negative, while in 27 patients Congored stain were positive for amyloid.

Conclusion: Regarding normal light microscopy in some patients with Amyloidosis, Electron microscopy is mandatory for diagnosis of early stage of amyloidosis in suspicious cases.
Study of the anxiety, stress, and depression in patients with chronic kidney disease undergoing hemodialysis in the Bandar Imam Khomeini Rah-Zainab Hospital in 2015

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Introduction: Chronic kidney disease (CKD) is a health problem and can lead to major changes in the patient's life style, which affects health status and quality of life. The present study aimed to evaluate the anxiety, stress and depression in patients with CKD undergoing hemodialysis in the Bandar Imam Khomeini Rah-Zainab Hospital.

Methods: The study was a descriptive-analytical study. Forty three patients undergoing hemodialysis in the Bandar Imam Khomeini Rah-Zainab Hospital were selected by simple random sampling method. Demographic questionnaire, anxiety, stress, and depression (DASS 21) standard questionnaire were used to collect their data, respectively. Patients were survived by their complete satisfaction. For data analysis, Pearson correlation coefficient and one way ANOVA statistical methods were used. The data were statistically analyzed by the program SPSS23.

Results: The mean age was 41.98 ± 9.71 years and 53% of them were male. Regarding anxiety, there was no anxiety in 34.9% of the patients, 20.39% low, 23.3% moderate, and 20.9% were diametrical. Depression was low in 44.2%, moderate in 16.3% and the rest were normal. There was no correlation between depression and gender (r=0.17, p=0.26). There was a significant relationship between depression and financial situation (P=0.03). Thirty percent of the patients experienced stressful situations (20% female).

Conclusion: Regarding relatively high rate of anxiety and depression in patients with CKD, appropriate interventions should be considered to improve their quality of life and health status. By providing care and supporting programs, family members and medical teamscan help them to cope with the disease.
Bardet-Biedl syndrome with end-stage renal disease: A case report of rare condition with congenital renal hypoplasia

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Introduction: End stage renal disease (ESRD) represents a clinical condition in which there is an irreversible loss of endogenous renal function. Bardet-Biedl syndrome (BBS) is a rare autosomal recessive disorder, characterized by clinical and genetic heterogeneity such as polydactyly, central obesity, mental retardation, hypogonadism, hypercholesterolemia, and vision loss. ESRD in BBS patients is the final stage of the disease and increases mortality of them. Our aim is to describe a 12 years old Iranian boy who had BBS.

Case report: The first presentation was fever and convulsions in age two. On physical examination the patient had polydactyly in both hands and both legs. Abdominal sonography showed renal hypoplasia. Undeveloped brain and cerebellum was showed in brain CT-scan. Other laboratory tests and physical examination were normal. At the age three he was hospitalized due to persistence high grade fever and seizure in the last few days. After several workups and based on his clinical presentation, the BBS was diagnosed. Also, mental retardation, respiratory distress, hypertension, difficulty in moving the eye, cerebral arachnoid cyst, reflux nephropathy and retarded bone age were diagnosed. After follow-up of the patient, at the age of 12, he was diagnosed with ESRD and hemodialysis was started for him.

Conclusion: Many associated minor features can be helpful in making a diagnosis and are important in the clinical management of BBS. Close follow-up for renal involvement in patients with BBS from an early age is highly recommended to postpone ESRD and so renal replacement therapy. This case exemplifies the need for multidisciplinary management in such cases.
The relationship between hemodialysis and dental caries and periodontal health in children

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Introduction: Chronic kidney disease (CKD) is an increasing public health issue. Prevalence is estimated to be 1.3-1.7% among Iranian Children. Despite the fact that the hemodialysis therapy have increased the survival rate among the patients affected by renal disease, dental health among the patients undergoing hemodialysis has been found to be debilitated and gets worsened with the increased duration of hemodialysis. Thus, this study aimed to assess the relationship between hemodialysis and dental caries and periodontal health in children with CKD.

Methods: This study was based on articles achieved from, Science direct, Pub Med, Medline, Scopus and Google Scholar in chronological order of 2010-2016 with using standard and proper key words (Hemodialysis, Dental health, Duration of dialysis, Kidney diseases, and periodontal disease). In primary search 138 articles were obtained, and after reviewing the title and abstract 35 studies were selected and the full texts were examined. Eighteen articles were cross sectional and only 8 articles contained inclusion criteria (The patients without the presence of an additional infectious disease, Patients without seizures or nervous disorders, patients without drug dependency) and they reached the final stage of research.

Results: Assessment of the 8 studies revealed that in patients undergoing hemodialysis VMI(mm/tooth), salivary urea, saliva pH and buffer capacity, plaque index (PI), probing pocket depth (PPD) are significantly higher, although the dental caries, decayed, missing, filled (DMF) were significantly lower.

Conclusion: Generally patients with CKD suffer from dental and periodontal disorders. Due to Insufficient enamel development and mineralization and salivary characteristic, oral hygiene and periodontal status are in a poor condition, but because of increase in salivary urea level, pH and buffer capacity of saliva, there was a significant decrease in dental caries experience.
Vitamin D3 supplementation reduces wet-nights in 7-15 years old children with nocturnal enuresis

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Introduction: Vitamin D deficiency is associated with nocturnal enuresis (NE) with an unknown mechanism. Recent studies showed that children with NE have low level of serum vitamin D and Vit D supplements decreased the incidence of NE. The renal action of vitamin D and its role in sleep disorder might be possible mechanism of Vit D for control of enuresis. In this study we examined the effect of vitamin D supplementation for treatment of NE in children.

Methods: Participants were 90 children (mean age: 8.5 y) with NE who were selected from children referred to pediatric clinic of Imam Reza, Shiraz. Children with NE were randomly assigned into two groups. The first group received vitamin D capsules (1000IU) and the second group received placebo for 2 months. Urine calcium/creatinine ratio and serum calcium were checked before the study. The number of wet-nights and the frequency of urination/night were reported each week during the study.

Results: There was 38 children in treatment group (22 boys and 16 girls) and 37 children in control group (21 boys and 16 girls). There was no gender difference between two groups. After two months, the mean number of wet-nights/week significantly decreased in treatment group in comparison with control group (p-value= 0.002). The reduction of the mean wet-nights/week was also significant in the intervention group before and after the study (p-value<0.001). The participants in treatment group had lower number of urination/night after supplementation and also lower numbers of urination in comparison with placebo group, too.

Conclusion: It was concluded that Vitamin D might be a safe therapy for NE but further studies are needed to determine the appropriate dose of Vit D for control of NE in children.
Prevalence of FTT in children with chronic kidney disease in Hazrat Ali Asghar Hospital, 1393-1394

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Introduction: Children with chronic kidney disease suffer from poor growth. The current study aimed to determine the prevalence of FTT in children with chronic kidney disease (CKD) in Hazrat Ali Asghar Hospital.

Methods: In this cross-sectional study, the population consisted of 113 children between 2 to 16 years with CKD who referred to Hazrat Ali Asghar Hospital from Khordad 1393 to Tir 1394. The inclusion criteria was children with CKD without other underlying diseases (heart, lung and metabolic disease). The independent variables were age, gender, family history of renal disease, and the age at the time of diagnosis of CKD. Dependent variables were body mass index, height, weight, MUAC, skin fold thickness, Gomez index, and the intensity and the kind of FTT. The obtained information entered into the data gathering form (check list) and finally analysis was done by SPSS version 18.

Results: One hundred thirteen children were examined. The mean age was 7.2 ± 4.4 years old. The 43.4% of the children were girls. The mean age at the time of diagnosis of CKD was 4.3 ± 3.3 years. The average of serum creatinine was 3.7 ± 3.1 mg/dl, and the average amount of GFR was 36.6 ± 34.3 ml/min/1.73m². Regarding the prevalence of FTT, 12.4% had mild FTT, 30.1% moderate FTT and 11.5% had severe FTT. A meaningful relationship was observed between FTT and GFR (p=0.0001) and among a group of children without FTT, the amount of GFR was higher. There was also a meaningful relationship between creatinine and FTT (p=0.0001) and among those without FTT the serum creatinine was lower. The meaningful relationship between gender and MUAC was not observed (p>0.05). There was no association between age, gender and FTT (p>0.05).

Conclusion: The results of the current study showed that half of the children with CKD had different degrees of growth retardation that was associated with the lower GFR.
Comparative evaluation of pre and post hemodialysis hemoglobin levels, their association with quality of life and the subsequent difference in erythropoietin dosage in patients with end-stage renal disease

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Introduction: Postdialysis hemoglobin (Hb) of hemodialysis (HD) patients is closer to real Hb level and can be a better reference for erythropoietin (EPO) dosage calculation. We measured the predialysis and postdialysis Hb concentrations of HD patients to calculate the decline in EPO dosage prescription and subsequent cost reduction using postdialysis Hb level as the reference. Additionally, the correlation between predialysis and postdialysis Hb and quality of life parameters was determined.

Methods: In this cross-sectional study, we measured predialysis and postdialysis Hb of 52 HD patients. The adjusted EPO dosage using predialysis and postdialysis Hb was calculated. We computed the EPO dosage decline and estimated the cost reduction. In addition, we calculated the correlation of all 11 items of SF-36 questionnaire with predialysis and postdialysis Hb.

Results: The mean Hb level rise after HD was 7.0±6.0%. Using the predialysis Hb, 34.6% of patients had a high Hb (>11.5g/dL); however, this percent increased to 55.8% using the postdialysis Hb. The mean required EPO according to postdialysis Hb would be significantly lower comparing with the predialysis Hb (10947±6820 vs 12047±7542 U/week, P<0.001). Thus, using postdialysis Hb for EPO dosage calculation causes significant cost reduction: 17.57±11.00 vs 15.96±9.85 dollars/patient/week for predialysis and postdialysis Hb, respectively (P<0.001). This action causes saving of 83.72 dollars/patient/year. None of the items of SF-36 questionnaire had significant correlation with predialysis and postdialysis Hb.

Conclusions: Using postdialysis Hb as the reference of EPO administration in HD patients results in significant reduction in EPO dosage and cost.
The Frequency of Kidney Stones in Terms of Composition in Patients Referring to Lithotripsy Center in Ilam, Western Iran

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Introduction: Kidney stone is a common clinical disorder and the prevalence rate in Iran is higher than the global mean. Ilam province is also one of the areas that are located on the so-called stone belt. The purpose of this study was to estimate the frequency of different types of kidney stones in Ilam city in order to adopt appropriate strategies for planning and preventing of this disease.

Methods: This descriptive analytical study was conducted on 170 patients referring to the Asia Lithotripsy Center in Ilam, Iran. Data were collected using bipartite questionnaire consisting of demographic data and type of stones. The first part was completed by interviewing the patients and the second part was filled after receiving the results of laboratory analysis of stones. Finally, obtained data was entered into SPSS version 16 and statistical analysis was performed.

Results: Complete analysis of the stones indicated that the prevalence of kidney stones was 68% in males and 31.8% in females with the M/F ratio of 2:1. There was no significant correlation between gender and type of stones (P=0.09). The highest prevalence of stones was between the ages of 31-41 (33.7%) and also a significant relationship was found between age and type of stones (P=0.00). The frequency of stones composition was: calcium oxalate (61.2%), mixed (36.2%), uric acid (62%), cystine (1.8%), respectively. Among mixed stones, calcium oxalate+uric acid (21.8%) and calcium oxalate+calcium phosphate (10.6%) stones had higher prevalence.

Conclusion: In the present study, the calcium oxalate and uric acid+calcium oxalate stones were more prevalent. Considering the high prevalence of these stones, preventive measures should be considered in addition to medical treatment to reduce the risk of kidney stones.
Urine evaluation in children with monosymptomatic nocturnal enuresis

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Introduction: Nocturnal enuresis (NE) is a common symptom in children. Most of the children with NE have no underlying organic cause. However, urine evaluations including urinalysis and urine culture is usually recommended. Therefore, we analyzed the results of urine evaluation in children presenting with monosymptomatic NE.

Methods: In this cross-sectional study, 147 consecutive children referring to a nephrology clinic with monosymptomatic nocturnal enuresis and no suspected underlying organic cause for enuresis were included. After history taking and physical examination, urinalysis and urine culture were performed.

Results: The patients had 6 to 14 years of age with mean age of 8.4 years. Male to female ratio was 1.5. The only positive urine culture was in a 6-year-old girl with 5-6 WBC/hpf in urinalysis. Abnormal findings of urinalysis included: pyuria (≥5 WBC/hpf) in 12 patients, hematuria (≥5 RBC/hpf) in 8 patients and proteinuria in 2 patients. No glucosuria was detected. All the samples had specific gravity of more than 1.014 except for one with 1.010.

Conclusion: We may conclude that in children with monosymptomatic NE and normal physical examination, urine culture may not be indicated unless with abnormal urinalysis.
تعیین میزان افت هموگلوبین بعد از عمل درون مجاری پروستات (TURP) در بیماران دارای فشار خون بالا و مقایسه آن با بیماران دارای فشار خون طبیعی

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سایه و هدف:
طی بروزتات، شایع تورم خوش خیم در مردان می‌باشد که بروز آن وابسته به سن است. عمل جراحی درون مجاری (TURP) یکی از روش‌های درمانی طلایی در راهی‌پیمایی خوش خیم بروزتات است. هدف از این مطالعه تعیین میزان افت هموگلوبین بعد از عمل جراحی درون مجاری پروستات (TURP) در بیماران دارای فشار خون بالا و مقایسه آن با بیماران دارای فشار خون طبیعی است.

مواد و روش ها:
گروه شاهد و مورد در این مطالعه شامل 40 نفر از بیمارانی هستند که با علائم بزرگ‌خور خیم بروزتات و بر اساس معیارهای مشخص داشته علائم سنجش ادراری تحتانی (LUTS) به بیمارستان وی‌وی‌سی (عج) مراجعه کرده‌اند. نمونه‌ها به دو گروه بیماران با فشار دارای فشار خون بالا و نرمال تقسیم شدند و یک سال دور بررسی قرار گرفتند. در این مطالعه هموگلوبین یکبار قبل از عمل و سپس روزه‌ای اندامه‌گیری هموگلوبین روز دوم پس از عمل به عنوان معیار افت هموگلوبین چاپ 1393

نتایج چگونه:
به راه‌هایی همیشه مشاهده شد که افزایش فشار خون بالا، نسبت به افراد دارای فشار خون نرمال بیشتر.

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New neurologic findings in a boy with schimke immuno-osseous dysplasia: A case report

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Introduction: Schimke immuno-osseous dysplasia (SIOD) is a rare autosomal recessive disease that caused by biallelic mutation in SMARCAL1 gene. Up to now, only three patient of SIOD was reported in Iran. Common findings in SIOD include; steroid resistance nephrotic syndrome due to focal segmental glomerulosclerosis (FSGS), progressive renal failure, spondyloepiphyseal dysplasia, cerebral infarction, and T-cell immunodeficiency. In this case report, we report a six year old boy with cerebral infarction and tremor of extremities.

Case report: The prob and was the second child of healthy consanguineous (3rd degree) parents. After birth, ventricular septal defect (VSD) and pulmonary hypertension was diagnosed and repaired. He had growth failure and recurrent respiratory infection in childhood. On physical examination, syndactyly of the second and third toes of the left foot, café au lait spots over the trunk, and right undescended testis were observed. After prescription of Rituximab at the age of 5.5 year old, he developed fever and oral aphthous lesion and 3 weeks later tremor of both hands and feet. Complete blood count test showed leukopenia. Triglyceride, cholesterol, LDL, and ESR were higher than the normal range. Brain MRI relieved a periventricular banding and capping in favor of micro vascular process. There were also bithalamic infarctions extending to the midbrain, infarction in the right inferior cerebellum, anda lacunar infarction in the left side deep frontal white matter. Brain MRA showed no arteriovenous malformation and no significant narrowing of arteries. Genetic MRA showed homozygote mutation of SMARCAL1 gene.

Conclusion: SIOD is a rare disease that commonly causes cerebral infarction, renal dysfunction and hematologic malignancy. Tremor of extremities should be considered as a new neurologic finding due to cerebral infarction in patients with SIOD.
A study on the rate, clinical features and etiology of urolithiasis in children younger than 15 years: Systematic Review

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Introduction: Urinary stones are among the most common complaints in patients referring to nephrologist and urologists. Although Urolithiasis occurs less often in children than adults, but the incidence is increasing and it causes considerable morbidity and hematuria may be the only presenting sign. Metabolic derangements, infection, neurogenic bladder and urinary obstruction are the major risk factors of urolithiasis. The aim of this study was to determine the prevalence, clinical signs and risk factors of urolithiasis in children younger than 15 years old.

Methods: In this systematic review study; cross-sectional articles published in databases including Iran Medex, SID, Medlib, PubMed, Scopus and Google scholar during the years 2002-2016 were reviewed. Keywords were "Urolithiasis", "Renal stone", "Children", "Hypercalciurian" and related words. All non-related studies were excluded.

Results: In most studies the prevalence of urolithiasis in children younger than 15 years was relatively low but in a study from Mofid hospital, Tehran, the authors concluded that renal stone is common in pediatric patients. In almost all of the studies hematuria was the most important clinical sign but fever and dysuria was the commonest clinical features in Ahwaz study. Metabolic disturbances was the most common cause of stone formation in children. hyperuricosuria was the most common metabolic finding instead of hypercalciuria.

Conclusion: It was concluded that all children with urolithiasis should be completely evaluated in terms of metabolic risk factors. Early diagnosis and management of renal stones in children is necessary to prevent the development of renal failure.
Childhood Enuresis in Traditional Persian Medicine

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Introduction: Enuresis is one of the most common problems in children and can cause many problems for mothers and also low quality of life in children. We decided to describe enuresis in children in traditional Persian medicine (TPM).

Methods: This is a review article, using sources of TPM, including Tebb-e-Akbari, zakhireh-Kharazmshahi, Alaghraz-altabiat-va-mabahes-alalaniat, Al-ghanoon fi-al-teb, ect. to describe this condition in children.

Results: According to this study, the most common cause of enuresis in children in TPM are coldness and weakness of the bladder. Therefore, the treatment of this condition in TPM is warming the bladder with hot drugs. One of the most effective drugs is rubbing hot oil such as Liliumledebouri and Foeniculumvulgareoil along with Boswellia, Cyperus, Quercus and Punicagranatum on the site of the bladder, inferior of the abdomen. Also TPM recommends Golghand (as one of traditional Persian drug component) as an effective drug, and eating dry foods, and Barbecueas the best food for these children and water limitation at night. Randomized clinical trials has revealed safety and efficacy of TPM advices in the treatment of enuresis. Good tolerability, lack of serious side-effects and drug interactions are the advantages of this preparations.

Conclusion: TPM recommends simple advices, which are affordable, available, safe and cheap with minimal side effects, rather than different medical treatments and interventions for children with enuresis.
The effect of erythropoietin against motor dysfunctions induced by bilateral renal ischemia in male rats

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Introduction: Neurologic sequelae remains a common and destructive problem in patients with acute kidney injury (AKI). The objective of the present study was to evaluate the possible neuroprotective effect of erythropoietin (EPO) on motor impairments following bilateral renal ischemia (BIR) in two time points after reperfusion: short term (24h) and long term (1w).

Methods: Male Wistar rats underwent BIR or sham surgery. EPO or saline administration was performed 30 min before surgery (1,000 IU/kg, i.p.). Explorative behaviors and motor function of the rats was evaluated by open field, rotarod and wire gripe tests.

Results: Plasma concentrations of blood urea nitrogen (BUN) and creatinine (Cr) were significantly enhanced in BIR rats 24h after reperfusion. BIR group had only an increased level of BUN but not Cr 1w after reperfusion. Impairment of balance function by BIR was not reversed by EPO 24h after reperfusion, but counteracted 7 days after ischemia. Muscle strength had no significant differences between the groups. BIR group had a decrease in locomotor activity and EPO could not reverse this reduction in both time point of the experiment.

Conclusion: Although EPO could not be offered as a potential neuroprotective agent in the treatment of motor dysfunctions induced by BIR, it could be effective against balance dysfunction 1w after ischemia.
Characterization of the Most Common Mutation in CTNS Gene in Iranian Patients with Infantile Nephropathic Cystinosis

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Introduction: 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 cells of different organs. Cystinosis has a worldwide incidence between 1:100000 to 1:200000 live births; its frequency has not been studied in Iranian population. The most common mutation in Northern European population is the 57-kb deletion. Reports proving the 57-kb deletion mutation has not been observed so far in any of the Middle East studies, including Egypt, Iran, Turkey and Saudi Arabia. The aim of this study was analysis of CTNS mutations in 20 Iranian patients with infantile nephropathic cystinosis from 20 unrelated families.

Methods: Mutation screening was performed by PCR amplification and sequencing of all 10 coding exons of CTNS gene in patients.

Results: Among 20 patients with infantile nephropathic cystinosis from 20 unrelated families that were participated in this study, a previously reported splice site mutation, c.681G>A; E227E, was detected in 11 patients in homozygous state and in 2 patients in heterozygous state.

Conclusion: Splice site mutation, c.681G>A, that comprises 60% (24 alleles) of the mutant alleles of all patients in this study, is the common mutation in the Middle East. According to former studies in the Middle East, this mutation is distributed with different frequencies in Iran (39.5%), Turkey (20%), Saudi Arabia (15.4%), and Egypt (7.7%) but has not been reported in none of the European and American populations up to now. Thus we suggest that c.681G>A is the most common mutation in Iran on the basis of this study and the previous study of cystinosis in southwestern of Iran and it can be called as a founder mutation in the Middle East with the maximum frequency in Iran.