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Case Report **Published Date:-2023-12-19 10:27:21**

Kidney Biopsy in Autosomal Dominant Polycystic Kidney Disease

Proteinuria is an easily quantified biomarker of kidney disease and often a sign of glomerular pathology. Significant proteinuria is uncommon in cystic kidney diseases and should be further evaluated to exclude the presence of another simultaneous kidney disease. While renal biopsy is a valuable part of the diagnostic evaluation of proteinuria, careful consideration of risks and benefits is necessary before proceeding in a patient with bilateral renal cysts. We report the case of a man with Polycystic Kidney Disease (PKD) who was found to have nephrotic-range proteinuria. An ultrasound-guided kidney biopsy revealed evidence of Focal Segmental Glomerulosclerosis (FSGS), which was attributed to hyperfiltration-related injury in the context of extensive kidney cysts. Genetic testing did not reveal a cause of FSGS and showed a variant of uncertain significance in PKD1. We use this case to highlight three important issues that are applicable to patients with PKD: the role of diagnostic evaluation for proteinuria in cystic kidney disease, the feasibility of kidney biopsy despite the presence of bilateral renal cysts, and the roles and limitations of genetic testing in cystic kidney disease and FSGS.

Case Report **Published Date:-2023-11-14 14:11:43**

Rare Case of Dense Deposition Disease with Combined C3 and C4d Deposits with MYH9-related Mutation

Background: The C3 glomerulopathies are a group of rare forms of glomerulonephritis with an incidence of 1-2 cases per million. It is mainly characterized by dysregulation of the alternative complement pathway. It is further classified morphologically based on electron microscopy ultrastructural findings into Dense Deposition Disease (DDD) and C3 glomerulonephritis. DDD is normally characterised by C3 Deposits.
Case: We report a rare case of a young Emirati male who presented with sub nephrotic proteinuria and microscopic haematuria on routine evaluation. Renal biopsy showed features of DDD with combined C3 and C4 deposits. The retinal evaluation showed features of Drusen classically seen in DDD. Genomic study showed heterozygous mutation in c.5842G>C (p.Asp1948His) variant of uncertain significance in MYH9 gene.
Discussion: C3 Glomerulopathy is a type of immune mediated disease previously classified as membranoproliferative glomerulonephritis. DDD is mainly characterised by C3 deposits in the glomerular basement Membrane. Our case has both C3 and C4d deposits, which is a rare entity. It shows the activation of both classical and alternate pathways.
Conclusion: Dense deposition disease is a rare complement mediated glomerulopathy. It is characterised by C3 deposits. Dense deposition disease with combined C3 and C4d deposits is a new entity. The treatment and prognosis of such cases will be different and unique compared to the normal cases of DDD.

Research Article **Published Date:-2023-10-20 15:31:55**

Effects of Zinc Supplementation on Oxidative Stress in Patients Undergoing Maintenance Hemodialysis

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Introduction: The aim of this study was to examine the effects of Zn supplementation on oxidative stress by evaluating changes in serum Copper (Cu) to Zinc (Zn) ratio, homocysteine (hCys), Glutathione (GSH), Total Bilirubin (TB) and Catalase (CAT) activity in hemodialysis patients.
Methods: Seventy-seven HD patients were enrolled in a multicenter simple-blind randomized clinical trial. Only 37 HD patients completed the study. They were randomly divided into two groups and supplemented with zinc sulfate (n = 17) or placebo (n = 20) for two months. Serum Zn and Cu were measured by atomic absorption spectrophotometry. Serum hCys was measured by immunology method, serum GSH and CAT activity were assessed by spectrophotometry method and TB was measured by colorimetric method. Determinations were performed before and after supplementation.
Findings: After zinc supplementation, serum Zn, serum GSH, and Serum Total Bilirubin (STB) significantly increased. Serum Cu to Zn ratio, serum hCys, and CAT activity significantly decreased in the Zn Zn-supplemented group.
Conclusion: Zinc supplementation increased serum antioxidant factors such as Zn, GSH, and bilirubin and decreased serum oxidative factors such as copper to zinc ratio, hCys, and decreased CAT activity. The study results suggest that zinc supplementation may be a useful tool for the improvement of oxidative stress in HD patients.

Research Article **Published Date:-2023-10-13 12:15:29**

Circulating Levels of Fibroblast Growth Factor 23 Selective for C-Terminal (FGF23-CT) in Hemodialysis Patients

Background: In hemodialysis patients, fibroblast growth factor 23 (FGF23) has reportedly been associated with the development of cardiovascular complications and a high risk of mortality. Our objective here was to study the cleavage characteristics of FGF23 in hemodialysis patients.
Methods: This study design is a cross-sectional observational investigation of three facilities without intervention. To assess FGF23 concentrations, we obtained plasma samples from 97 hemodialysis patients before the hemodialysis session and from 16 healthy volunteers. We measured the FGF23 C-terminal fragment and intact FGF23 concentrations by using a commercial enzyme-linked immunosorbent assay.
Results: Serum levels of the FGF23 C-terminal fragment were 189 ± 121 ng/mL in healthy volunteers and 306 ± 206 ng/mL in hemodialysis patients. The ratios of intact FGF23 to total FGF23 were 0.03 ± 0.03 in healthy volunteers and 0.44 ± 0.28 in hemodialysis patients. The ratios were positively correlated with levels of inorganic phosphate in hemodialysis patients ($p < 0.001$, $r = 0.52$).
Conclusion: We measured actual levels of the serum FGF23 C-terminal fragment in hemodialysis patients by using a new commercial kit for the first time. The ratio of intact FGF23 to total FGF23 was lower in healthy controls than the ratio in hemodialysis patients. The cleavage percentage of FGF23 was considerably higher in both groups of subjects than previously thought. We suggest that hyperphosphatemia in hemodialysis patients was associated with impaired cleavage of FGF23.

Research Article **Published Date:-2023-10-10 12:41:32**

Clinical and Epidemiological Profile of Reversible Acute Kidney Injury with Full Recovery: Experience of a Nephrology Department

Purpose: Acute kidney injury (AKI) is a real public health problem due to its severity and gravity. In a 2013 meta-analysis, Susantitaphong, et al. estimated the incidence of AKI worldwide at between 10% and 20%. In the latter study, no African studies were included, given the lack of data in the literature. Our objective was to identify the clinical and paraclinical epidemiological characteristics of patients with AKI.
Patients and methods: We conducted a retrospective study including patients who had AKI with recovery of normal renal function hospitalized in a nephrology service between 2002 and 2015.
Results: Our population consisted of 107 men and 107 women with a median age of 61 years (IQR 43-73.25) of which 42.1% were multitargeted. Functional AKI represented the predominant mechanism of AKI retained in our study with a rate of 53.2% with dehydration as the main etiology for 108 patients (50.46%). Organic cause was retained in 38.8% of patients, with acute tubular necrosis (ATN) as the most frequent etiology (37.35%). Kidney disease improving global outcomes (KDIGO) stage 3 was the stage retained for 115 patients included in our series, 31 of whom required extra renal purification. During their hospitalization, 78.5% of the patients presented a persistent AKI (duration of the episode > 7 days). A glomerular filtration rate (GFR) lower than 60 ml/min/1.73 m^2 at discharge was found in 119 patients and 10 patients had a GFR higher than 90 ml/min/1.73 m^2. After 3 months from discharge, 77.5% of patients had a GFR between 60 and 90 ml/min/1.73 m^2.
Conclusion: Our results give us an idea of the epidemiological and clinical characteristics of patients who have had acute renal failure with recovery of normal renal function and enable us to better recognize patients at risk in order to avoid such complications. AKI remains a major issue and knowing its epidemiological and clinical characteristics will allow its prevention.

Case Report **Published Date:-2023-10-05 16:29:25**

A Case of Catastrophic Atypical Hemolytic Uremic Syndrome Unresponsive to Eculizumab and the use of Ravulizumab Off-label

“A 40-year-old woman with melanoma, under treatment with Dabrafenib and Trametinib, was evaluated in our hospital for rapidly progressive deterioration of renal function”.
8 months before the current admission, the patient had been diagnosed with melanoma, and underwent radical surgery and subsequent therapy with Dabrafenib and Trametinib.
After 5 months of therapy, the patient was brought to this hospital for precordial pain, with a diagnosis of myopericarditis, therapy was started for heart failure with a good response. However, the patient developed a progressive impairment of renal function, associated with hemolytic anemia and thrombocytopenia. The peripheral smear showed the presence of schistocytes.
The suspicion of atypical Hemolytic Uremic Syndrome (aHUS) was confirmed by the assay of C5B-9 induced by serum on endothelial cells, which showed a deposition of 331%, treatment with Eculizumab was initiated.
After 3 administrations the patient did not improve, with further worsening of the hemolytic condition, and progression of renal damage.
Due to the failure of Eculizumab, we considered the use of Ravulizumab. However, in Italy only can be administered to patients in Eculizumab stable treatment for at least three months. Nevertheless, faced with the catastrophic condition, it was decided to shift the therapy and use off-label Ravulizumab. After 10 days of the first administration, the laboratory tests showed a continuous rise in the values of haptoglobin, platelets, and hemoglobin, and a decrease in LDH. The renal function failed to return to normal values but after 20 days of therapy with Ravulizumab, there was complete resolution of the hemolytic condition.

Research Article **Published Date:-2023-09-29 00:00:00**

Doppler Evaluation of Renal Vessels in Pediatric Patients with Relapse and Remission in Different Categories of Nephrotic Syndrome

Aim: To study resistivity & pulsatility indices in the interlobar arteries of kidneys in patients with idiopathic nephrotic syndrome and evaluate their response to steroids
Method: The prospective case-control study was carried out in 100 patients, aged 2 years - 15 years of nephrotic syndrome and divided into 5 groups depending upon their response to steroids. Twenty-five age-matched controls were included for comparison. The abdominal ultrasonography and Doppler examinations were performed for patients of each group and for the controls.
Result: The mean Resistivity Index (RI) and Pulsatility Index (PI) of the interlobar arteries were calculated for the five subgroups of nephrotic syndrome patients and for the controls. There was a significant difference in mean RI in all the evaluated arteries between the 6 groups. No significant difference was noted in the pulsatality indices of the kidney of the cases of nephrotic syndrome when compared to controls.
Conclusion: Steroid-resistant nephrotic syndrome patients show a statistically significant difference in RI and not a statistically significant difference in PI Doppler parameters when groups of patients compared to controls. These parameters may be used to predict the response to steroids.
