Introduction: Accumulation of ammonia in the blood may result in an acute life-threatening event in children. Patients typically present with lethargy, poor feeding, and hypotonia; if not managed efficiently, patients can develop seizures, coma, and eventually die. Management of hyperammonemia is difficult in children given the non-specific symptoms, age-specific etiologies, and lack of consensus in the treatment plan. We systematically reviewed published literature to provide expert consensus panel recommendations for medical management and renal replacement therapy (RRT) in pediatric patients with hyperammonemia.

**Methods:** PubMed/Medline, Embase and Cochrane database search was performed to include studies about hyperammonemia and RRT in children <18 years old. Two independent reviewers reviewed each title, abstract, and relevant full text articles. An expert panel of international pediatric nephrologists discussed medical management and RRT for hyperammonemia in children at a consensus conference to provide recommendations.

**Results:** The initial search returned a total of 477 citations and 25 studies met the inclusion criteria. A total of 132 patients were included in these 25 studies and were treated with different dialysis modalities. Twenty-three hyperammonemia patients were treated with peritoneal dialysis with 65% success rate, five were treated with intermittent hemodialysis with 100% success rate, 92 were treated with continuous renal replacement therapy (CRRT) with 60% success rate and three were treated by extracorporeal membrane oxygenation (ECMO) combined with CRRT and had 100% success rate.

**Conclusions:** Expert panel recommendations were provided with regards to non-RRT (medical management), hemodialysis, peritoneal dialysis, CRRT, high dose CRRT, and hybrid therapy. CRRT was the first line dialysis modality of choice recommended for hyperammonemia management in children. Indications for RRT were variable among studies reviewed; RRT was recommended at ammonia blood level > 400mmol/L or in hemodynamically unstable children irrespective of blood ammonia level. More studies are needed to further strengthen these expert recommendations.

**SAT-315**

**ROLE OF P-GLYCO PROTEIN /MULTIDRUG RESISTANCE-ASSOCIATED PROTEIN-1 ON DIFFERENT T-C细粒体 SUBSETS AND EFFECT OF MDR-1 GENE POLYMORPHISM ON P-GP EXPRESSION IN IDIOPATHIC NEPHROTIC SYNDROME IN CHILDREN**

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**Introduction:** Glucocorticoids remains mainstay of therapy for Idiopathic Nephrotic Syndrome (INS). Other than histological changes, pharmacogenomics factors may also affect steroid response. Overexpression of P-glycoprotein (P-gp) and Multidrug resistance associated protein 1 (MRP-1) modulate the pharmacokinetics of steroids and may contribute in steroid resistance.

**Methods:** P-gp, MRP-1 expression were evaluated on whole blood and functional activity on peripheral blood mononuclear cells (PBMCs) in steroid sensitive nephrotic syndrome (SSNS) (n=170, male 103, mean age=8.54±4.3); steroid resistant nephrotic syndrome (SRNS) (n=81, male 43, mean age=7.43±4.6) patients. The genetic variants G2677T/A (rs2032382) of MDR-1 gene was genotyped using polymerase chain reaction-restriction fragment length polymorphism (PCR-RFLP) technique.

**Results:** Demographic significant difference were found in 24hrs urinary protein/creatinine ratio [SSNS=0.13±0.06, SRNS=3.67±0.91, p<0.001], total cholesterol [SSNS=144.21±34.61, SRNS=460.52±201.09, p<0.001], triglyceride [SSNS=135.08±36.20, SRNS=413.44±171.78, p<0.001]. Percentage of P-gp (10.35±1.25 v/s 4.19±1.07, p=0.001); and MRP-1 (17.03±3.45 v/s 8.71±0.97, p<0.001) positivity on PBMC were significantly higher in SSNS than SRNS. P-gp expression on CD4+ (6.08±1.06 v/s 4.34±1.47, p=0.008); and CD8+ cells (8.65±2.19 v/s 3.99±1.72, p<0.001) significantly high in SSNS than SRNS respectively. MRP-1 expression on CD4+ and CD8+cells significantly higher in SSNS (12.06±2.91 v/s 3.35±0.83, p=0.043); (5.11±0.68 v/s 1.59±0.39, p<0.001) respectively. Functional activity of P-gp and MRP-1 was significantly increased in SSNS as compared to SRNS (66.12±12.71 v/s 28.22±7.35, p<0.001); (72.30±8.38 v/s 32.38±8.89, p<0.001) respectively. With ROC curve analysis of 81 steroid resistant patients’ predictive cut-off values for the percentage of P-gp and MRP-1 was found to be 6.99% and 9.64 % respectively with sensitivity of 95% and 90.1% and specificity of 99.4% and 90.6% respectively (Figure 1). Moreover homozygous mutant allele TT+AA was significantly associated with resistant population of nephrotic syndrome (p=0.025, OR = 2.86 CI=1.14-7.14). However heterozygous (GT+GA) and mutant alleles (T+A) were not significantly associated with steroid resistant nephrotic syndrome patients as compared to steroid sensitive nephrotic syndrome patients.

In our data the set expression of P-gp (7.50±3.79 v/s 5.65±3.22, p=0.016) was significantly higher in the patients of homozygous mutant alleles compared to wildtype GG (Figure 2).

**Conclusions:** Overexpression of P-gp and MRP-1 on CD4+ and CD8+ T cells contribute in resistance to corticosteroids and polymorphism of variants G2677T/A may promote P-gp expression in steroid resistant in INS in children . Use of P-gp and MRP-1 inhibitors may prevent SRNS status.

**SAT-316**

**HEALTH RELATED QUALITY OF LIFE OF CHILDREN WITH NEPHROTIC SYNDROME IN LAGOS NIGERIA**

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**Introduction:** Nephrotic syndrome is the leading cause of renal disorders in Africa and accounts for more than 50% of cases seen in our setting. The characteristic chronic relapsing nature and the response to steroid have significant implications on outcome. The quality of life of children with this disorder in Africa have not been extensively studied. The aim of this study was to assess the quality of health of children with nephrotic syndrome and factors affecting them.

**Methods:** A cross sectional study of children to assess the quality of life in children aged 2years to 18 years with idiopathic nephrotic syndrome using the Pediatric Quality of Life Inventory (PedsQL TM 4.0 Generic Core Scales) was conducted. This was conducted between Jan 2018-June 2018. Clinical and demographic details were recorded. Data obtained for physical, emotional, social and school domains were analyzed using the Statistical Package for Social Sciences (SPSS) version 19.0.

**Results:** Sixty-one children were recruited, males 37(60.7%) and females 24(39.3%). Mean age at first diagnosis in years was 5.16(SD 3.39) and a mean duration of illness of 83.26 weeks (SD 91.62 ). 49(84.5%) had one or more relapses in six months or a year, 26(52%) had no relapse. The mean PedsQL score difference based on steroids (p=0.002) and school functioning domains were significant (p= 0.012 and 0.010 respectively) and the total mean difference was significant with p= 0.012. There was no significant difference in the mean PedsQL score based on duration of illness except in the parent assessment with respect to school functioning (p=0.016). Based on their steroid sensitivity, mean comparison of scores for parent and child showed significance in the physical, emotional and social functioning and overall for parent’s assessment (p < 0.05) and physical functioning domain in child assessment (p <0.05). The mean PedsQL score difference based on chronic kidney disease (CKD) and estimated GFR showed in the child assessment, significant impact in emotional functioning (p= 0.026 and 0.0009 respectively).

**Conclusions:** The overall health quality of life of children with Nephrotic syndrome in our setting is good. However, those who were not steroid sensitive NS, with CKD and longer duration of illness had poorer quality of life.

**SAT-317**

**THE IMPACT OF CARING FOR CHILDREN WITH POSTERIOR URETHRAL VALVES**

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