genetic patients, there was CR to ciclosporin in 36.5% (19/52), to tacrolimus in 33.3% (9/27) and to cyclophosphamide in 11.5% (3/26).
2. Biopsy findings showed no correlation with either outcome, or response to IIS
3. 87.5% of patients with genetic disease showed no or partial response to IIS
3. In patients testing negative for genetic disease, those with secondary steroid resistance had a significantly higher response rate than those with primary steroid resistance (41.5 vs 21.8%), and the highest response rate was to Rituximab (66.7%)
4. Strikingly, 93% of patients who responded to 1st IIS showed no progression (ERF), after up to 10 years of follow-up.
5. 74.3% of non-responders progressed to established renal failure (ERF) within the follow up period, with 50% recurrence rate post-transplant
6. 50% of Rituximab non-responders relapsed post-transplant, and remain resistant to therapy
Conclusions: This study stratifies non-genetic patients into responders and non-responders to IIS, with markedly different outcomes, and strongly suggests two distinct underlying immune mechanisms.

P-317 EVALUATION OF PLATELET INDICES AS NOVEL MARKERS FOR NEPHROTIC SYNDROME

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Introduction: Platelets play an important role in the pathogenesis of nephrotic syndrome (NS). The aim of this study was to evaluate platelet indices as markers for this disorder.
Material and methods: A total of 39 patients (26 males, 13 females) with NS followed-up by the pediatric nephrology department in our center from January 2017 to January 2018 were enrolled in the study. Participants were divided into two groups, those with steroid-sensitive nephrotic syndrome (SSNS) (n = 23) and those with steroid-resistant nephrotic syndrome (SRNS) (n = 13). Laboratory parameters, including neutrophils, lymphocytes, white blood cells, platelet counts, mean platelet volume, platelet distribution width, plateletcrit, and platelet large cell ratio were reviewed retrospectively. Similar parameters from a control group consisting of 30 age- and sex-matched healthy subjects were also evaluated and compared to the study groups.
Results: White blood cells, neutrophil count, platelet, and plateletcrit values were greater in NS patients when compared to controls. When evaluated based upon steroid resistance status, no statistical difference was observed between the SSNS group and controls in terms of platelet indices; however, plateletcrit values were greater in the SRNS group compared to the control group ($P = 0.007$).
Conclusions: Plateletcrit could be useful as a marker of inflammation for SRNS. More studies with large sample sizes should be performed to determine changes in platelet indices that occur in association with NS.

P-318 NEPHROTIC SYNDROME IN THE FIRST YEAR OF LIFE: CASE SERIES OF 28 INFANTS

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Introduction: Nephrotic syndrome (NS) with onset within 1st year has a varied etiology and prognosis. **Aim:** To study clinical spectrum, histopathological findings and outcome of infantile NS. **Design:** Retrospective observational study.
Material and methods: Twenty eight infants with NS with onset within one year of life (Infantile NS) were subclassified as Congenital NS (3 cases) and Infantile NS (25 cases). Case records were reviewed for clinical features, investigations including TORCH infections, clinical course and complications. Renal Biopsy was done were indicated.
Results: Fifty seven percent of cases were between 9-12 months of age. Male: female ratio was 1.33:1. Associated abnormalities were seen in 9 cases (32%). Cases of congenital nephrotic syndrome had overall poorer outcome with only one of 3 cases survived, having normal renal function at last follow-up. Of the 25 cases of infantile nephrotic syndrome, 19 cases had normal renal function at last follow-up, 4 cases progressed to chronic kidney disease and 2 cases were lost to follow-up.
Renal biopsy was done in 14 of 25 cases of infantile NS. Most common histopathological finding seen was Minimal change Nephrotic syndrome (MCNS)(6) followed by Focal segmental glomerular sclerosis (FSGS) (4), congenital nephrotic syndrome Finnish variety (2) and Diffuse Mesangeal sclerosis (1).
Conclusions: "Finnish variety" was common in congenital NS and MCNS was found to be the most common histopathological finding in the Infantile variety of nephrotic syndrome. Associated anomalies are frequently associated. Significant number (52%) of cases responded to Steroids.

P-319 TARGETED NEXT GENERATION SEQUENCING IN CHIDREN WITH INFANTILE NEPHROTIC SYNDROME

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Introduction: Infantile nephrotic syndrome (NS) is a rare, genetically heterogeneous group of glomerulopathies with the onset of the disease at the age of 4-12 months. We aimed to determine the causative gene mutations in children with infantile NS through next generation sequencing (NGS).
Material and methods: The mutational analysis was performed in 8 children (4M/4F) aged 3.2 (IQR: 3.0; 6.1) years with infantile NS from nonconsanguineous families. Renal biopsy revealed FSGS in 7/8 (87.5%) the affected children. Targeted NGS covering 68 genes associated with steroid-resistant NS (SRNS) with confirmation by direct Sanger sequencing were applied.
Results: A total of 13 different mutations were detected in 9/68 (13.2%) genes associated with SRNS in 7/8 (87.5%) children with infantile NS. 3/13 (23.1%) of these mutations were novel. Monogenic cause of infantile NS was identified in 4/8 (50%) children. We determined homozygous mutations in *NPHS2* gene (n=2), compound-heterozygous mutations in *NPHS1* and *ITGB4* genes (n=1), heterozygous mutations in *WT1* gene (n=1). Digenic inheritance had 2 patients: *PTPRO* with *SCARB2* (n=1) and *WT1* with *ITGB4* (n=1). Extrarenal syndromic features were presented in 3 of the affected children: Wilms tumor (n=2), microcephaly with deafness (n=1). 2 patients with homozygous *NPHS2* mutations progressed to ESKD by the age of 6 years. Kidney transplantation was performed in one of them without disease recurrence after transplantation for 14 months.
Conclusions: We conclude that monogenic cause of infantile NS was identified in 50% of children with the most prevalent mutations in *NPHS2*, *NPHS1*, *ITGB4* and *WT1* genes. A molecular genetic diagnosis of infantile NS through NGS may have important consequences for the clinical management of patients and prediction of a risk for disease recurrence after kidney transplantation.

P-320 ISOLATED PROTEINURIA DUE TO FOCAL SEGMENTAL GLOMERULOSCLEROSIS ASSOCIATED WITH MUTATIONS IN THE LAMA5 GENE IN A BOY

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