

# Clinical & Pediatric Nephrology

May 09-10, 2016 New Orleans, USA

## Clinical, treatment and prognosis in children with FSGS: A 5 year retrospective study

Ling Chen, Liu Xiaorong, Shen Ying, Chen Zhi, Fan Jianfeng and Jiang Yeping  
Capital Medical University, China

**Aim:** To investigate the clinical and pathological characteristics in children with Focal segmental glomerular sclerosis and to evaluate the associated factors of clinical outcome and the efficacy of treatment.

**Methods:** The clinical and pathological characteristics, the associated factors of clinical outcome and the efficacy of treatment were analyzed retrospectively in 62 children with FSGS from June 2007 to June 2012.

**Results & Conclusion:** Based on clinical and pathological classification: 49 patients with edema and proteinuria (79.0%), 25 patients with hematuria (40.3%), 15 patients with hypertension (24.2%), 7 patients with acute renal injury (9.6%). Renal pathology revealed 45 patients with not otherwise specified type (NOS) (72.6%), 9 with glomerular tip lesion type (Tips) (14.5%), 8 with cellular type (Cellular) (12.9%). The complete remission rate of NOS was 28.9% (13/45) and the partial remission rate was 33.3% (15/45) and no remission rate was 37.8% (17/35). The complete remission rate of Tips was 45.6% (5/9) and the partial remission rate was 33.3% (3/9) and no remission rate was 11.1% (1/9). The complete remission rate of Cellular was 25% (2/8) and no remission rate was 75% (6/8). The prognosis correlated with the pathological characteristics and the therapy plan ( $r=0.142, 0.327$ , respectively,  $P<0.05$ ). Of these FSGS patients, 12% patient experienced remission after Prednisone and cyclophosphamide pulse therapy (P+CTX) and the total efficacy was 40%, which was statistically lower than that of children who received Prednisone and Cyclosporin A (P+CsA) or Prednisone and Tacrolimus (P+TAC) (37.5% and 75% or 42.9% and 71.4%, respectively). No significant statistical difference in prognosis was found in patients with P+CsA or P+TAC group ( $p>0.05$ ). High blood pressure and persistent large proteinuria do harmful for the prognosis ( $P<0.05$ ) rather than age, gender, pathological type and treatment plan.

[15210938018@163.com](mailto:15210938018@163.com)

## Genetic analysis of Chinese childhood steroid resistant nephrotic syndrome

Jie Ding  
Peking University First Hospital, China

Genetic steroid resistant nephrotic syndrome caused by at least 30 genes is an important cause of end stage renal disease. However, there is a scarcity of data for the frequency of single gene causes of childhood steroid resistant nephrotic syndrome in large cohorts of Chinese patients using targeting next generation sequencing. Using a multi-gene next generation sequencing panel of nephrotic syndrome related 28 genes, we performed genetic analysis of 120 patients from 5 centers in China with steroid resistant nephrotic syndrome manifested before 18 years of age. Disease causing mutations for ten genes were detected in 34/120 patients with the mutation detection rate of 28.3%. Of the 120 patients, 8 patients had ADCK4 mutations (6.67%), 7 patients had NPHS1 mutations (5.83%), 7 patients had WT1 mutations (5.83%), 4 patients had NPHS2 mutations (3.33%) and 8 patients had other 6 genes mutations together (6.67%). A de novo mutation in TRPC6 was detected in a patient with infantile onset nephrotic syndrome. The age of onset of ADCK4 nephropathy ranged from 1 month to 17 years. In conclusion, ADCK4 was the most frequent causative gene in Chinese childhood steroid resistant nephrotic syndrome and the mutation detection rate of NPHS2 was lower than that of literature data.

[djnc\\_5855@126.com](mailto:djnc_5855@126.com)