

#1933 - The effect of parental consanguinity on clinical course and outcome of children with focal segmental glomerulosclerosis; a report from Iran.

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Body

Background: FSGS is responsible for 20% of nephrotic syndrome in children and 75% of steroid resistant nephrotic syndrome. The main aim of this study was to evaluate the effect of parental consanguinity on clinical course and outcome of children with FSGS.

Methods: This historical cohort study was conducted on 69 children with steroid resistant FSGS. Clinical characteristics at baseline and last evaluation and response to treatments were recorded. History of parental consanguinity was obtained in all patients.

Results: Forty four patients (63.8 %) were male and male to female ratio was 1.76:1. The mean age at disease presentation was 5.69 ± 2.39 (range: 1-10) years. Fifty one patients (73.5 %) were born form consanguineous parents. The resistance rate to cyclosporine A and cyclophosphamide were significantly higher in patients with parental consanguinity than without it. The mean renal survival time was significantly lower in patients with consanguinity than without consanguinity (8.33 vs. 10.44 years, $P=0.02$). In univariate analysis, parental consanguinity was a risk factor for progression to chronic kidney disease (CKD) (HR=4.56, 95% CI 1.06-19.47, $P=0.04$).

Conclusions: FSGS patients with parental consanguinity had lower renal survival time, higher resistance rates to treatments, and also higher risk of progression to CKD.

References

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