

ASK THE EXPERT

A 10-year-old girl was found to have asymptomatic proteinuria upon routine urinary screening at school. The subsequent renal biopsy revealed focal segmental glomerulosclerosis. The girl had nephrotic-range proteinuria without nephrotic syndrome. There was no microhematuria. Her creatinine clearance and blood pressure were also normal. How should this girl be managed?

Key words Primary focal and segmental glomerular sclerosis · Management

This young girl has persistent proteinuria without complete nephrotic syndrome, normal blood pressure, and normal creatinine clearance. The term primary focal and segmental glomerular sclerosis (FSGS) applies to this disease, where glomeruli not involved by segmental lesions have no histological or ultrastructural abnormalities.

It is unlikely that the child has a reduced renal mass. An association between primary FSGS and renal hypoplasia was described long ago, but in such cases creatinine clearance is usually decreased. However, renal ultrasonography should be performed to measure the size of the kidneys. It is also assumed that another secondary form of FSGS, such as human immunodeficiency virus infection, glycogen storage disease, reflux nephropathy, or Alport syndrome, has been excluded.

Patients with primary FSGS without nephrotic syndrome most often, in our experience, do not respond to corticosteroid therapy and also fail to respond to alkylating agents. According to McAdams et al. [1], only one-quarter of these patients progress to end-stage renal disease. There seems to be no risk of recurrence in the graft. Hence we do not recommend an aggressive course of steroids and/or alkylating agents. There are no data on the efficacy of cyclosporine in this setting. Furthermore, it has not been proven whether induction of remission of proteinuria by any form of therapy prevents progression to renal failure [2].

We would rather favor the use of angiotensin converting enzyme (ACE) inhibitors. Although the efficacy of these drugs has not been demonstrated in patients with primary FSGS, studies in adult patients have shown that ACE inhibitors may reduce proteinuria and prevent progression to renal failure in secondary forms of FSGS due to diabetes or IgA nephropathy [3, 4]. The side effects of treatment with ACE inhibitors are minimal compared with those observed with steroids and alkylating agents.

The editors invite questions for this section

References

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LITERATURE ABSTRACT

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Changing trends of histopathology in childhood nephrotic syndrome

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This study was conducted to analyze the trend of histopathologic subtypes in idiopathic nephrotic syndrome (INS) in a homogenous racial group in India population. A prospective analysis of 400 consecutive children with INS was performed. Kidney biopsies were performed according to standard indications. Steroids were administered following the Arbeitsgeminschaft für Padiatrische Nephrologie protocol. Cyclophosphamide was administered to children in the frequent-relapser, steroid-dependent, and steroid-nonresponder categories. Of the various histopathologic subtypes, focal segmental glomerulosclerosis (FSGS) was the most common (87 of 222 subtypes; 39.1%). Children who underwent biopsy between July 1992 and December 1996 (group B, n = 157) were compared with our initial published data of biopsies performed between January 1990 and June 1992 (group A, n = 65), with similar indications for biopsy in both groups. The incidence of FSGS was significantly greater in biopsies performed in the recent period (group B, 47% versus group A, 20%; P = 0.0002). The different clinical and biochemical parameters were also analyzed to differentiate FSGS from the other 2 subtypes. Hypertension (P = 0.005), renal insufficiency at presentation (P = 0.001), and steroid resistance (P = 0.0006) were significantly greater in children with FSGS. On follow-up (mean, 5.4 years), children with FSGS were at a significantly greater risk for developing renal insufficiency (P = 0.0001). We conclude there is a shift toward an increasing prevalence of FSGS over the years in the Indian population. This trend has immense therapeutic and prognostic significance.