

Focal segmental glomerulosclerosis: use of other therapies

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GUIDELINES

No recommendations possible based on Level I or II evidence

SUGGESTIONS FOR CLINICAL CARE

(Suggestions are based on Level III and IV evidence)

There have been a number of case series using mycophenolate mofetil in patients with resistant focal segmental glomerulosclerosis (FSGS). Most demonstrate that although mycophenolate mofetil can induce some reduction of proteinuria, complete remission of proteinuria is rare. No data on long-term follow-up evaluation with this drug are currently available.

- **Cattran et al (2004)** performed an open-label, 6-month trial of mycophenolate mofetil in 18 patients with biopsy-proven FSGS who were resistant to corticosteroid therapy. Seventy-five per cent had also failed to respond to a cytotoxic agent and/or a cyclosporin. A substantial improvement in proteinuria was seen in 44% (8/18) of patients by 6 months. However, no patient achieved complete remission. In addition, relapses were common after therapy was discontinued.
- **Briggs et al (1998)** previously reported the use of mycophenolate mofetil in 7 patients, in whom a substantial improvement in proteinuria was also observed.
- **Gellermann et al (2004)** investigated the effect of mycophenolate mofetil in 7 children with a resistant nephrotic syndrome (6 of whom had minimal change disease and one with FSGS). In this patient, mycophenolate mofetil resulted in complete remission for a follow-up of 28 months.

Other therapies have been used in patients with FSGS who prove resistant to standard treatment:

- **Partial remission** has been observed in a few case reports using tacrolimus (McCauley et al 1990).
- **Vincristine** has also been used for the treatment of steroid- and cyclophosphamide-resistant nephrotic syndrome. In a series of eight cases presented by Goonasekera et al (1998), two children treated with vincristine achieved complete remission associated with preserved renal

function. Another experienced transient relapses. Although studied in primary FSGS, there may be particular advantages of vincristine in secondary forms of nephrotic syndrome associated with malignancy (see Guideline titled “FSGS: cytotoxic therapy”).

- **Plasma exchange, lipid apheresis and immunoadsorption have also been reported to induce remission of proteinuria in selected patients (Hattori et al 2003, Haas et al 1998, Mitwalli 1998).**

Background

Despite the use of steroids, cytotoxic therapy, and cyclosporine, some patients with idiopathic FSGS are unable to establish or maintain sustained clinical remission of proteinuria and progress inexorably toward end-stage kidney disease (ESKD). The objective of this guideline is to evaluate the available clinical evidence pertaining to the impact of interventions not covered in other guidelines on renal functional decline in patients with idiopathic FSGS.

Search strategy

Databases searched: MeSH terms and text words for focal segmental glomerulosclerosis were combined with MeSH terms and text words for mycophenolate mofetil, tacrolimus, vincristine, plasma exchange, lipid apheresis, immunoadsorption and other therapies. This search was carried out in Medline (1966 to September Week 2, 2004). The Cochrane Renal Group Trials Register was also searched for trials not indexed in Medline.

Date of searches: 17 September 2004.

What is the evidence?

There have been no randomized controlled trials (RCTs) of these additional agents.

Summary of the evidence

There is currently insufficient evidence for any specific benefit from other therapies in the treatment of resistant nephrotic syndrome due to focal and segmental glomerulosclerosis.

What do the other guidelines say?

Kidney Disease Outcomes Quality Initiative: No recommendation.

UK Renal Association: No recommendation.

Canadian Society of Nephrology: No recommendation.

European Best Practice Guidelines: No recommendation.

International Guidelines: No recommendation.

Implementation and audit

No recommendation.

Suggestions for future research

No recommendation.

References

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