



Complete Summary

GUIDELINE TITLE

Focal segmental glomerulosclerosis: use of cyclosporine A.

BIBLIOGRAPHIC SOURCE(S)

Thomas M. Focal segmental glomerulosclerosis: use of cyclosporin A. Westmead NSW (Australia): CARI - Caring for Australasians with Renal Impairment; 2005 Sep. 9 p. [9 references]

Thomas M. Focal segmental glomerulosclerosis: use of cyclosporine A. Nephrology 2006 Apr;11(S1):S185-8.

GUIDELINE STATUS

This is the current release of the guideline.

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SCOPE

DISEASE/CONDITION(S)

Idiopathic focal segmental glomerulosclerosis

GUIDELINE CATEGORY

Management
Treatment

CLINICAL SPECIALTY

Family Practice
Internal Medicine

Nephrology
Pediatrics

INTENDED USERS

Physicians

GUIDELINE OBJECTIVE(S)

To evaluate the available clinical evidence pertaining to the impact of cyclosporin A on renal functional decline in patients with idiopathic focal segmental glomerulosclerosis

TARGET POPULATION

Adults and children with idiopathic focal segmental glomerulosclerosis:

- Steroid resistant
- Steroid dependent
- Frequent relapse

INTERVENTIONS AND PRACTICES CONSIDERED

Treatment

Cyclosporin A therapy

MAJOR OUTCOMES CONSIDERED

- Renal function
- Remission rate

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Databases searched: MeSH terms and text words for focal segmental glomerulosclerosis were combined with MeSH terms and text words for cyclosporin A therapy. This search was carried out in Medline (1996 to September Week 2, 2004). The Cochrane Renal Group Trials Register was also searched for trials in focal segmental glomerulosclerosis not indexed in Medline.

Date of searches: 17 September 2004.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Levels of Evidence

Level I: Evidence obtained from a systematic review of all relevant randomized controlled trials (RCTs)

Level II: Evidence obtained from at least one properly designed RCT

Level III: Evidence obtained from well-designed pseudo-randomized controlled trials (alternate allocation or some other method); comparative studies with concurrent controls and allocation not randomized, cohort studies, case-control studies, interrupted time series with a control group; comparative studies with historical control, two or more single arm studies, interrupted time series without a parallel control group

Level IV: Evidence obtained from case series, either post-test or pretest/post-test

METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review with Evidence Tables

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

Not stated

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not applicable

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

Comparison with Guidelines from Other Groups
Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

Recommendations of Others. Recommendations regarding treatment of focal segmental glomerulosclerosis with cyclosporin A from the following groups were discussed: Kidney Disease Outcomes Quality Initiative, UK Renal Association, Canadian Society of Nephrology, European Best Practice Guidelines, and International Guidelines.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Definitions for the levels of evidence (I–IV) can be found at the end of the "Major Recommendations" field.

Guidelines

Cyclosporin may be effective in preserving filtration function in patients with steroid-resistant focal segmental glomerulosclerosis (FSGS), in those with steroid dependence or in those who frequently relapse on conventional therapy. (Level II evidence)

Suggestions for Clinical Care

(Suggestions are based on Level III and IV evidence)

A number of open studies have shown that cyclosporin is able to induce complete and partial remission in both adults and children with steroid resistant FSGS and steroid-dependent FSGS. Partial or complete remission is most likely in steroid-dependent FSGS, while the response rate in steroid-resistant FSGS is variable, ranging between 20% and 70% in most studies.

Cyclosporin is also associated with significant toxicity, which means that use of this agent should be reasonably restricted to patients at high risk of end-stage kidney disease (ESRD), or in whom toxicity from steroid-dependence confers a greater danger than chronic cyclosporin therapy.

What Dose Should Be Used?

Optimal dosing and monitoring of cyclosporin has not been fully clarified. Most studies have used doses of approximately 5 mg/kg/day with a blood level of 100–200 mg/mL. (Level III evidence)

Should Steroids Also Be Used?

Most studies have also continued a low dose of steroids while using cyclosporin. There is anecdotal evidence that this approach may be more effective in achieving remission in children than cyclosporin alone.

Optimal Duration of Therapy

A minimum effective dose of cyclosporin should be continued for at least 2 years. (Level IV evidence) Relapse is common after tapering or discontinuing the drug. Patients who are in complete remission for more than 1 year on cyclosporin appear to be more likely to remain in remission if the cyclosporine is gradually tapered and discontinued, rather than stopped suddenly. (Level IV evidence, anecdotal reports)

Definitions:

Levels of Evidence

Level I: Evidence obtained from a systematic review of all relevant randomized controlled trials (RCTs)

Level II: Evidence obtained from at least one properly designed RCT

Level III: Evidence obtained from well-designed pseudo-randomized controlled trials (alternate allocation or some other method); comparative studies with concurrent controls and allocation not randomized, cohort studies, case-control studies, interrupted time series with a control group; comparative studies with historical control, two or more single arm studies, interrupted time series without a parallel control group

Level IV: Evidence obtained from case series, either post-test or pretest/post-test

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is identified and graded for each recommendation (see "Major Recommendations").

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

Appropriate use of cyclosporin A in the management of patients with focal segmental glomerulosclerosis

POTENTIAL HARMS

Cyclosporin is also associated with significant toxicity, which means that use of this agent should be reasonably restricted to patients at high risk of end-stage kidney disease, or in whom toxicity from steroid-dependence confers a greater danger than chronic cyclosporin therapy.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Living with Illness

IOM DOMAIN

Effectiveness

IDENTIFYING INFORMATION AND AVAILABILITY

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ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2006 Apr

GUIDELINE DEVELOPER(S)

Caring for Australasians with Renal Impairment - Disease Specific Society

SOURCE(S) OF FUNDING

Industry-sponsored funding administered through Kidney Health Australia

GUIDELINE COMMITTEE

Not stated

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Author: Merlin Thomas

FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

All guideline writers are required to fill out a declaration of conflict of interest.

GUIDELINE STATUS

This is the current release of the guideline.

GUIDELINE AVAILABILITY

Electronic copies: Available in Portable Document Format (PDF) from the [Caring for Australasians with Renal Impairment Web site](#).

Print copies: Available from Caring for Australasians with Renal Impairment, Locked Bag 4001, Centre for Kidney Research, Westmead NSW, Australia 2145

AVAILABILITY OF COMPANION DOCUMENTS

The following is available:

- The CARI guidelines. A guide for writers. Caring for Australasians with Renal Impairment. 2006 May. 6 p.

Electronic copies: Available from the [Caring for Australasians with Renal Impairment \(CARI\) Web site](#).

PATIENT RESOURCES

None available

NGC STATUS

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