

GUIDELINES FOR THE DIAGNOSIS AND MANAGEMENT OF RECURRENT FSGS IN PEDIATRIC TRANSPLANT RECIPIENTS

I. PURPOSE

To outline guidelines for the diagnosis and management of recurrent FSGS following renal transplantation in children.

II. SETTING

In-patient and out-patient

III. DEFINITIONS

Recurrent FSGS is diagnosed if any of the following are seen post-tx:

- Nephrotic range proteinuria: >3.5 g/24 hr or UPC >3 g/g in adults and FMV or 24 hr urine protein/Cr ratio of > 2g/g or >3+ on dipstick in children AND hypoalbuminemia of < 3 g/dL
- Allograft biopsy shows FSGS pattern or injury and widespread effacement of podocytes
- Early recurrence occurs within 48 hr; intermediate in 2-30 days, late recurrence may be after 3 months post-tx.
- Risk factors include progression to ESRD within 3 yr after dx of FSGS or rFSGS in prior tx

IV. GUIDELINES

Evaluation:

- Monitor urine protein and serum Cr daily for 1 week; twice a week in week 2, weekly for 4 weeks, monthly for the first year and every 3 months thereafter; preferably using the FMV sample.

Treatment of rFSGS:

- Prophylactic TPE or rituximab pre-tx is NOT recommended
- Prompt initial therapy with intensive TPE should be started - daily for 3 days then three times a week for 2 weeks. TPE can be stopped after reduction of proteinuria to < 1 g/d or UPC <1.
- ☐ Consider monitoring IgG levels and IVIG if infectious complications
- Alternatively Immunoabsorption may be used; 2.5-3 plasma volumes daily x 1 week followed by QOD for 2 weeks, then BIW for 2 more weeks.
- TPE or IA should also be used for relapse after stopping above Rx.
- CNI may be changed to CsA with high target levels (2 hr peak range 1200-1400 ng/ml until remission)
- Steroids may be used to enhance effect
- Rituximab should be considered; TPE should be held for 48 hr post rituximab dose to prevent drug removal.
- Consider conversion of MMF to oral Cytosin for 3 months
- LDL-A may be used in pts refractory to TPE or IA

IV. PROCEDURE

Write procedure here (an ideal series of steps that should be followed in a regular, definite order to fulfill policy)

VI. REFERENCES

Post-transplant recurrence of focal segmental glomerular sclerosis: consensus statements; Rupesh Raina, Swathi Jothi, Dieter Haffner, Michael Somers, Guido Filler, Prabhav Vasistha, Ronith Chakraborty, Ron Shapiro, Parmjeet S. Randhawa, Rulan Parekh et al: Kidney International, Volume 105, Issue 3, March 2024, Pages 450-463

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