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Efficacy of LDL Apheresis in Therapy Refractory Recurrent FSGS

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Introduction: FSGS (focal segmental glomerulosclerosis) is one of the idiopathic diseases that can causes nephrotic syndrome in the pediatric population. Secondary FSGS can be caused by a genetic mutation or other disease process. However, primary FSGS can occur without any known cause. Most pediatric patients present to their PCP or health center with unusual edema in face and lower extremities. Patients often describe their urine having a “foamy” appearance. Further initial work up show abnormal labs such an increase in serum creatinine, low albumin, and high cholesterol. Urine studies can show proteinuria and hematuria. Diagnosis is confirmed with a renal biopsy. However, genetic testing is important to determine treatment course.

The standard treatment for nephrotic syndrome consists of steroids, ACE inhibitors/ARBs, diuretics, and apheresis. This is a chronic disease process with the risk of relapse. However, some patients can develop refractory FSGS which eventually leads to renal failure.

Transplant can be challenging for those who have primary FSGS due to the risk of recurrence. Given the limited treatment options, there is a poor prognosis for recurrence post-transplant. Careful consideration regarding transplant as well as treatment plans both before and after transplant must be determined for each individual patient based on their disease process.

FSGS recurrence after kidney transplantation has been described in up to 50% of some pediatric cohorts. Decreased graft survival occurs in transplanted children with FSGS compared to other primary diagnoses in both living and deceased donor transplants, largely related to recurrence. Strategies to decrease FSGS recurrence have relied on plasmapheresis and intensification of immunosuppression, though some children remain refractory to these interventions. LDL apheresis has been proposed as a potential efficacious therapy in FSGS recurrence, though data in transplanted children is limited.

Methods: We present the cases of two children with recurrent FSGS after deceased donor kidney transplants. Both children had demonstrated FSGS resistance to conventional apheresis therapy as well as to marked intensification of immunosuppression. Both children started a 12-treatment trial of LDL apheresis over 9 weeks (2 sessions/week x 3 weeks and then weekly x 6 weeks) in the setting of nephrotic syndrome and significant graft dysfunction. Baseline immunosuppression was maintained during LDL apheresis. Clinical parameters assessing response to treatment were assessed over the LDA apheresis course.

Results: Both children tolerated LDL apheresis without sequelae. Both manifested resolution of edema and partial remission of FSGS with significant improvement in graft function. Both children maintained improved clinical parameters as LDL apheresis was tapered.

Conclusion: We conclude that: 1) LDL apheresis may be a useful adjunctive therapy in recurrent FSGS refractory to usual maneuvers; 2) Although only achieving a partial remission of FSGS, volume balance as assessed by weight and clinical exam improved significantly; 3) Graft function similarly improved significantly; 4) Clinical improvement persisted as LDL apheresis was weaned.

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