CHALLENGES OF MONITORING AND TREATING RECURRENT OF PRIMARY FOCAL SEGMENTAL GLOMERULOSCLEROSIS AFTER RENAL TRANSPLANTATION

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Background
Introduction In patients under 16 presenting with nephrotic syndrome, primary focal segmental glomerulosclerosis (FSGS) is an important differential. High recurrence rates following transplantation, and a considerable risk of progression to end-stage renal disease, are well-recognised in cases with presumed circulating permeability factors rather than a genetic cause. To limit the risk of graft loss, it is therefore crucial to monitor for relapsing disease. The main marker of recurrence is proteinuria, however this can be challenging to interpret in a patient with high proteinuria pre-transplantation.

Objectives This is a case of a 14-year-old with FSGS, who developed early recurrence post-transplantation.

Methods
Case Presentation Pre-emptive renal transplantation was discussed with the family of a 14-year-old male with steroid-resistant FSGS, who had also not responded to tacrolimus, mycophenolate and rituximab. Proteinuria would be difficult to interpret post-transplantation, as he continued to produce high levels of proteinuria with a significant urine output. A number of options were explored including; (1) bilateral nephrectomies or embolization pre-transplantation to shut down native function altogether; (2) a tube into the transplant renal pelvis passing through the bladder, serving as a catheter to monitor urine exclusively from the transplant kidney; (3) delay of transplantation until native kidney function completely ceased. Given the challenging practicality of (2), and uncertain timescale of (3), bilateral nephrectomies were arranged a few weeks prior to transplantation.

Results
Outcome After day five following a live donor kidney transplant, proteinuria developed progressively. Daily therapeutic plasma exchange was commenced for two weeks. Renal function continued to deteriorate, and lipoprotein apheresis was trialled. A transplant renal biopsy, nine days following lipoprotein apheresis, reported early recurrence of FSGS.

Conclusions

Discussion The leading cause of renal graft failure in primary FSGS, within the paediatric cohort, is disease recurrence. Removal of circulating permeability factors using extracorporeal systems, such as therapeutic plasma exchange and lipoprotein apheresis, could theoretically improve graft survival. Despite a number of possibilities considered, this case highlights the difficulties of monitoring and treating disease recurrence post-transplantation.