

INF2

Charcot-Marie-Tooth, FSGS

Mode of Inheritance	<ul style="list-style-type: none">• Autosomal dominant
Renal Phenotype	<ul style="list-style-type: none">• Proteinuria, end-stage renal disease• Age of onset: Childhood and young adulthood• Typical biopsy findings: FSGS
Extra-renal Manifestations	<ul style="list-style-type: none">• Sequelae from nephrotic syndrome:<ul style="list-style-type: none">• Hypoalbuminemia and edema• Hyperlipidemia• Hypothyroidism• Loss of immunoglobulins• Peripheral neuropathy
Pre-Transplant Management	<ul style="list-style-type: none">• Avoidance of steroid and intensive immunosuppression therapy• Avoidance of renal biopsy• Screening for and management of extra-renal manifestations
Transplant Considerations	<ul style="list-style-type: none">• Tailor immunosuppression given lower risk of recurrence post-transplant• Careful screening of potential living related donors (inherited in an autosomal dominant manner)
Post-Transplant Management	<ul style="list-style-type: none">• Lower risk of disease recurrence (4.5% vs 28.5%) (Trautmann <i>CJASN</i> 10:592, 2015)