

WT1

Denys-Drash Syndrome, Frasier Syndrome

Mode of Inheritance	<ul style="list-style-type: none">• Autosomal dominant• Most cases are sporadic
Renal Phenotype	<ul style="list-style-type: none">• Steroid resistant nephrotic syndrome, nephropathy, Wilm's tumor• Age of onset: Infancy and childhood• Typical biopsy findings: FSGS, DMS
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• Gonadoblastoma• XY gonadal dysgenesis• Ambiguous genitalia
Pre-Transplant Management	<ul style="list-style-type: none">• Avoidance of steroid and intensive immunosuppression therapy• Avoidance of renal biopsy• Consider prophylactic gonadectomy
Transplant Considerations	<ul style="list-style-type: none">• Tailor immunosuppression given lower risk of recurrence post-transplant
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)