

# PLCE1

## Nephrotic Syndrome

<b>Mode of Inheritance</b>	<ul style="list-style-type: none"><li>• Autosomal recessive</li></ul>
<b>Renal Phenotype</b>	<ul style="list-style-type: none"><li>• Steroid resistant nephrotic syndrome</li><li>• Age of onset: Infancy or childhood</li><li>• Typical biopsy findings: DMS (more common), FSGS</li></ul>
<b>Extra-renal Manifestations</b>	<ul style="list-style-type: none"><li>• Sequelae from nephrotic syndrome:<ul style="list-style-type: none"><li>• Hypoalbuminemia and edema</li><li>• Hyperlipidemia</li><li>• Hypothyroidism</li><li>• Loss of immunoglobulins</li></ul></li></ul>
<b>Pre-Transplant Management</b>	<ul style="list-style-type: none"><li>• Rare reports of children who do respond to steroid treatment (Hinkes <i>Ped Nephrol</i> 23:847, 2008)</li><li>• Avoidance of renal biopsy</li></ul>
<b>Transplant Considerations</b>	<ul style="list-style-type: none"><li>• Tailor immunosuppression given low risk of recurrence post-transplant</li></ul>
<b>Post-Transplant Management</b>	<ul style="list-style-type: none"><li>• Lower risk of disease recurrence (4.5% vs 28.5%) (Trautmann <i>CJASN</i> 10:592, 2015)</li></ul>