

COL4A5

Alport Syndrome

Mode of Inheritance	<ul style="list-style-type: none">• X-linked dominant
Renal Phenotype	<ul style="list-style-type: none">• Hematuria, proteinuria• Age of onset:<ul style="list-style-type: none">• Affected males: childhood• Carrier females: later onset, usually in 2nd or 3rd decade of life
Extra-renal Manifestations	<ul style="list-style-type: none">• Cataracts, myopia, lens opacities• Sensorineural deafness
Pre-Transplant Management	<ul style="list-style-type: none">• ACE inhibitors can delay the progression to ESRD
Transplant Considerations	<ul style="list-style-type: none">• Careful screening of potential living related donors (especially asymptomatic mothers who may develop disease later in life)
Post-Transplant Management	<ul style="list-style-type: none">• Low risk of disease recurrence if combined liver-kidney transplant• Approximately 3-4% of patients develop anti-GBM nephritis post-transplant (Kashtan <i>Medicine</i> 78:5, 1999)