PKHD1

Autosomal Recessive Polycystic Kidney Disease

Mode of Inheritance	• Autosomal recessive
Renal Phenotype	• Enlarged, cystic kidneys • Age of onset: Infancy or early childhood
Extra-renal Manifestations	• Liver fibrosis, portal hypertension, and hepatic cysts • Pancreatic cysts
Pre-Transplant Management	• Screening and management of extra-renal manifestations
Transplant Considerations	• Tailor immunosuppression given low risk of recurrence post-transplant • Consideration of a combined liver-kidney transplant
Post-Transplant Management	• Low risk of disease recurrence • Avoidance of hepatotoxic medications when possible

https://www.omim.org/entry/263200