HNF1B
Renal Cysts and Diabetes Syndrome

Mode of Inheritance	• Autosomal dominant • Variable expressivity and incomplete penetrance
Renal Phenotype	• Variable, including: renal cysts, hypodysplasia, interstitial fibrosis, and various other structural abnormalities
Extra-renal Manifestations	• Mature-onset diabetes of youth (MODY) • Subclinical abnormal LFTs • Neonatal cholestasis • Pancreatic hypoplasia • Genital tract abnormalities • Hyperuricemia and early-onset gout • Hypomagnesemia • Autism, ADHD, and developmental delay
Pre-Transplant Management	• Screening and management of extra-renal manifestations
Transplant Considerations	• Careful screening of potential living related donors (inherited in an autosomal dominant manner)
Post-Transplant Management	• Low risk of disease recurrence • Minimize exposure to pro-diabetogenic medications

OMIM: https://www.omim.org/entry/137920