## HNF1B

## **Renal Cysts and Diabetes Syndrome**

Mode of	Autosomal dominant
Inheritance	Variable expressivity and incomplete penetrance
Renal Phenotype	Variable, including: renal cysts, hypodysplasia, interstitial fibrosis, and various other structural abnormalities
	<ul> <li>Mature-onset diabetes of youth (MODY)</li> <li>Subclinical abnormal LFTs</li> <li>Neonatal cholestasis</li> </ul>
Extra-renal	
Manifestations	<ul> <li>Pancreatic hypoplasia</li> <li>Genital tract abnormalities</li> </ul>
Mannestations	
	Hyperuricemia and early-onset gout
	Hypomagnesemia     Autiom ADUD and developmental delay.
	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	<ul> <li>Low risk of disease recurrence</li> <li>Minimize exposure to pro-diabetogenic medications</li> </ul>

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