# Renal-hepatic-pancreatic dysplasia

INSERM

## Source

*INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Renal-hepatic-pancreatic dysplasia. ORPHA:294415*

Renal-hepatic-pancreatic dysplasia is a rare, genetic, developmental defect during embryogenesis syndrome characterized by the triad of pancreatic fibrosis (and cysts, with a reduction of parenchymal tissue), renal dysplasia (with peripheral cortical cysts, primitive collecting ducts, glomerular cysts and metaplastic cartilage) and hepatic dysgenesis (enlarged portal areas containing numerous elongated binary profiles with a tendancy to perilobular fibrosis). Situs abnormalities, skeletal anomalies and anencephaly have also been associated. Patients that survive the neonatal period present renal insufficiency, chronic jaundice and insulin-dependent diabetes.