

# Autosomalna recesywna wielotorbielowatość nerek

## Kod Orpha: 731 Kod OMIM: 617610

### Opis choroby \*

#### Definicja

A rare, genetic hepatorenal fibrocystic syndrome characterized by cystic dilatation and ectasia of renal collecting tubules, and a ductal plate malformation of the liver resulting in congenital hepatic fibrosis. Clinical presentation, whilst typically in utero or at birth, is variable and in the most severe cases includes Potter-sequence, oligohydramnios, pulmonary hypoplasia, and massively enlarged echogenic kidneys.

#### Dane

#### Klasyfikacja

Choroba

#### Synonimy

AR-PKD

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#### Kod ORPHA

731

#### Kod OMIM

617610

#### Kod ICD10

Q61.1

#### Kod ICD11

GB81

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[\\*Źródło](#)

orphanet

### Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.