

## Opis choroby \*

### Definicja

A rare, lethal, genetic, multiple congenital anomaly disorder characterized by the triad of brain malformation (mainly occipital encephalocele), large polycystic kidneys, and polydactyly, as well as associated abnormalities that may include cleft lip/palate, cardiac and genital anomalies, central nervous system (CNS) malformations, liver fibrosis, and bone dysplasia.

### Dane

<b>Klasyfikacja</b>	<b>Synonimy</b>
Zespół wad wrodzonych	Dysencephalia splanchnocystica Zespół Meckela i Grubera Meckel-Gruber syndrome

<b>Kod ORPHA</b>	<b>Kod OMIM</b>	<b>Kod ICD10</b>
564	616258	Q61.9

**Kod ICD11**  
LD2F.13

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### \*Źródło

orphanet