

## Opis choroby \*

### Definicja

A rare genetic lethal multiple congenital anomalies/dysmorphic syndrome characterized by severe hydranencephaly and renal dysplasia or agenesis. Pregnancy is complicated by oligo- or anhydramnios, leading to features of Potter sequence (including typical facies and microretrognathia, limb contractures, talipes equinovarus, and pulmonary hypoplasia) in the fetus. Affected fetuses either die *in utero* or shortly after birth. Histology of the brain shows widespread presence of multinucleated neurons and glial cells.

### Dane

<b>Klasyfikacja</b>	<b>Synonimy</b>
Zespół wad wrodzonych	MARCH syndrome
	MARCH syndrome

<b>Kod ORPHA</b>	<b>Kod OMIM</b>	<b>Kod ICD10</b>
500135	236500	Q87.8

**Kod ICD11**

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### \*Źródło

orphanet