

Wrodzone zespolenie wrotno-układowe

Kod Orpha: 480531 Kod OMIM:

Opis choroby *

Definicja

Congenital portosystemic shunt is a rare, congenital anomaly of the great veins characterized by an abnormal communication between one or more veins of the portal and the caval systems, resulting in complete or partial diversion of the portal blood away from the liver to the systemic circulation. Clinical manifestations include liver atrophy, hypergalactosemia without uridine diphosphate enzyme deficiency, hyperammonemia, encephalopathy (resulting in learning disabilities, extreme fatigability and seizures), pulmonary hypertension, hypoxemia from hepatopulmonary syndrome and benign or malignant tumours.

Dane

Klasyfikacja

Wada morfologiczna

Synonimy

Congenital portosystemic venous fistula
Wrodzona przetoka wrotno-układowa

Kod ORPHA

480531

Kod OMIM

-

Kod ICD10

Q26.5

Kod ICD11

DB98.Y

[*Źródło](#)

orphanet

Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.