Opis choroby *

Definicja

A rare renal tumor characterized by a unilateral, solitary, well demarcated, mesenchymal/myofibroblastic neoplasm occurring in very young children. Histopathologically, three subtypes (classic, cellular, and mixed) can be distinguished. The tumor most commonly involves the renal sinus and is typically discovered as a palpable abdominal mass. Patients may also present with hypertension or hematuria, rarely with hypercalcemia or hyperreninemia. Prenatal presentation, usually with polyhydramnios, is not infrequent. The most important prognostic factor is completeness of surgical resection. Overall, malignant potential is low and clinical outcome favorable.

Dane

Klasyfikacja
Choroba

Kod ORPHA
2665

Kod OMIM
-

Kod ICD10
D41.0

Kod ICD11
2C90.Y

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*Źródło

orphanet