Opis choroby *

Definicja

A rare subtype of congenital pulmonary airway malformation characterized by a multicystic mass of non-functioning lung tissue, consisting of small cysts of less than 2 cm in diameter. The lesions have intracystic communications, can be connected to the tracheobronchial tree, and are usually unilateral, involving a single lobe. The condition often presents with respiratory distress in the neonatal period or in infancy. It is frequently associated with other severe congenital anomalies, such as renal agenesis or dysgenesis, pulmonary sequestration, or cardiac abnormalities.

Dane

Klasyfikacja
Podtyp kliniczny

Synonimy
CCAM type 2
CCAM typu 2
CPAM typu 2
Wrodzona choroba torbielowata płuc typu 2
Wrodzona torbielowatość gruczolakowata płuc typu 2
CPAM type 2
Congenital cystic adenomatoid malformation of the lung type 2
Congenital cystic adenomatous malformation of the lung type 2
Congenital cystic disease of the lung type 2

Kod ORPHA
280840

Kod OMIM
-

Kod ICD10
Q33.0

Kod ICD11
LA75.4

---

*Źródło

orphanet