Opis choroby *

Definicja

A rare subtype of congenital pulmonary airway malformation characterized by a multicystic mass of non-functioning lung tissue consisting of numerous microcysts of less than 0.5 cm in diameter. The lesions have intracystic communications, can be connected to the tracheobronchial tree, and are usually unilateral, involving an entire lobe. The condition may be associated with polyhydramnios, fetal hydrops, and stillbirth, or present with severe respiratory distress in the neonatal period.

Dane

Klasyfikacja
Podtyp kliniczny

Synonimy
CCAM type 3
CCAM typu 3
CPAM typu 3
Wrodzona choroba torbielowata płuc typu 3
Wrodzona torbielowatość gruczolakowata płuc typu 3
CPAM type 3
Congenital cystic adenomatoid malformation of the lung type 3
Congenital cystic adenomatous malformation of the lung type 3
Congenital cystic disease of the lung type 3

Kod ORPHA
280847

Kod OMIM
-

Kod ICD10
Q33.0

Kod ICD11
LA75.4

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*Źródło

orphanet