Opis choroby *

Definicja

A rare subtype of congenital pulmonary airway malformation characterized by a multicystic mass of non-functioning lung tissue, with peripheral, large, thin-walled, often multiloculated cysts, which may be 8 cm in diameter. The lesions have intracystic communications, can be connected to the tracheobronchial tree, and are usually unilateral, involving a single lobe. Patients present with respiratory distress or respiratory infections in the neonatal period or in infancy. The condition is often associated with tension pneumothorax, signs of mediastinal shift, and malignant transformation to pleuropulmonary blastoma type 1.

Dane

Klasyfikacja
Podtyp kliniczny

Synonimy
CPAM type 4
CPAM typu 4
Wrodzona torbielowatość gruczolakowata płuc typu 4
Congenital cystic adenomatoid malformation of the lung type 4
Congenital cystic adenomatous malformation of the lung type 4

Kod ORPHA
280854

Kod OMIM
-

Kod ICD10
Q33.0

Kod ICD11
LA75.4

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*Źródło

orphanet