Opis choroby *

Definicja

A rare subtype of congenital pulmonary airway malformation characterized by global arrest of lung development with small, solid appearing lungs with a diffusely granular surface, histologically featuring bronchus-like structures with smooth muscle, glands, and numerous cartilage plates, embedded in loose, vascular mesenchymal tissue. The condition presents at birth and is incompatible with life.

Dane

Klasyfikacja
Podtyp kliniczny

Synonimy
CPAM type 0
CPAM typu 0
Wrodzona torbielowatość gruczolakowata płuc typu 0
Congenital cystic adenomatoid malformation of the lung type 0
Congenital cystic adenomatous malformation of the lung type 0

Kod ORPHA
280827

Kod OMIM
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Kod ICD10
Q33.0

Kod ICD11
LA75.4

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*Źródło

orphanet