

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

*Źródło

orphanet