

Opis choroby *

Definicja

A rare, non-syndromic respiratory or mediastinal malformation characterized by unilateral complete absence of lung tissue, bronchi, and pulmonary vessels. It may be isolated or associated with congenital malformations, most commonly with heart anomalies. Presentation is highly variable including airway narrowing, stridor, respiratory distress, recurrent respiratory tract infections, and pulmonary hypertension.

Dane

Klasyfikacja

Wada morfologiczna

Kod ORPHA

984

Kod OMIM

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Kod ICD10

Q33.3

Kod ICD11

LA75.1

*Źródło

orphanet