Opis choroby *

Definicja

A rare, genetic interstitial lung disease characterized by accumulation of lipoproteins in the pulmonary alveoli leading to restrictive lung disease and respiratory failure. Patients present with dyspnea, tachypnea, cough, failure to thrive, and digital clubbing. Liver disease have been described in some cases including hepatomegaly, steatosis, fibrosis or cirrhosis.

Dane

Klasyfikacja
Choroba

Synonimy
Hereditary pulmonary alveolar proteinosis with hepatic involvement
PAP, typ wyspy Reunion
Płucna proteinoza pęcherzykowa, typ wyspy Reunion
Śródmiąższowa choroba płuc i wątroby
Interstitial lung and liver disease
PAP, Reunion island type
Pulmonary alveolar proteinosis, Reunion island type

Kod ORPHA
440427

Kod OMIM
615486

Kod ICD10
J84.0

Kod ICD11
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*Źródło

orphanet