Opis choroby *

Definicja

A rare primary interstitial lung disease characterized by the accumulation of lipids and proteins related to surfactant in the alveoli in association with the presence of antibodies against granulocyte-macrophage colony-stimulating factor (GM-CSF). The disease leads to a progressive impairment of gas exchange and respiratory insufficiency.

Dane

Klasyfikacja
Choroba

Synonimy
Autoimmune PAP
aPAP
Autoimmunologiczna PAP
Idiopatyczna PAP
Idiopatyczna proteinoza pęcherzyków płucnych
iPAP
aPAP

Kod ORPHA
747

Kod OMIM
610910

Kod ICD10
J84.0

Kod ICD11
CB04.31

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*Źródło

orphanet