

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

*Źródło

orphanet