

Opis choroby *

Definicja

A rare idiopathic interstitial pneumonia characterized by prominent subpleural and parenchymal fibroelastosis and pleural fibrosis, predominantly involving the upper lobes. Signs and symptoms include non-productive cough, dyspnea, and recurrent respiratory infections. Pneumothorax is a frequently reported complication. Pulmonary function test reveals a restrictive pattern and reduced diffusing capacity. Computed tomography shows pleural thickening with signs of fibrosis (traction bronchiectasis, architectural distortion, and loss of volume), and reticulation.

Dane

Klasyfikacja

Choroba

Synonimy

IPPF

Idiopatyczna fibroelastoza opłucnej i mięszu płuca

Idiopathic pleuropulmonary fibroelastosis

Kod ORPHA

494428

Kod OMIM

-

Kod ICD10

J84.1

Kod ICD11

-

*Źródło

orphanet