

Opis choroby *

Definicja

A rare idiopathic interstitial pneumonia characterized by extensive, diffuse intra-alveolar accumulation of pigment-laden macrophages, most commonly associated with long-term exposure to tobacco smoke. Patients present with slowly progressive shortness of breath on exertion and chronic cough with bilateral crackles. Digital clubbing is also frequently observed. Pulmonary function test reveals a restrictive pattern. Computed tomography typically shows diffuse ground-glass opacities with subpleural and lower zone predominance.

Dane

Klasyfikacja

Choroba

Kod ORPHA

98852

Kod OMIM

263000

Kod ICD10

J84.1

Kod ICD11

CB03.3

*Źródło

orphanet