Opis choroby *

Definicja

A rare systemic autoimmune disease characterized by an aggressive fibroinflammatory process with infiltration of IgG4-positive plasma cells in the mediastinum, potentially resulting in compression and functional impairment of vital mediastinal structures, and associated with elevated serum IgG4. Clinical symptoms are unspecific and include pain or symptoms due to mass effect. The condition may occur together with IgG4-related disease in other parts of the body.

Dane

Klasyfikacja
Podtyp kliniczny

Synonimy
Fibrosing mediastinitis
Stwardnienie śródpiersia
Mediastinal fibrosis
Sclerosing mediastinitis

Kod ORPHA
63999

Kod OMIM
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Kod ICD10
J98.5

Kod ICD11
CB22.0

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*Źródło

orphanet