

Opis choroby *

Definicja

A rare systemic disease characterized by acute or subacute onset of thrombocytopenia, anasarca (edema, pleural effusion, ascites), and systemic inflammation (fever and/or elevated C-reactive protein). Minor diagnostic categories are Castleman's disease-like features on lymph node biopsy, reticulin myelofibrosis and/or increased number of megakaryocytes in bone marrow, progressive renal insufficiency, and mild organomegaly including hepatosplenomegaly and lymphadenopathy. Most patients show elevated levels of serum alkaline phosphatase, while marked polyclonal hypergammopathy is rare.

Dane

Klasyfikacja

Choroba

Synonimy

Thrombocytopenia-anasarca-fever-renal
insufficiency-organomegaly syndrome
Zespół małopłytkowości, uogólnionego obrzęku
skóry i tkanki podskórnej, gorączki,
niewydolności nerek i organomegalii

Kod ORPHA

457077

Kod OMIM

-

Kod ICD10

M35.8

Kod ICD11

4A4Y

*Źródło

orphanet