

Opis choroby *

Definicja

A rare lymphatic system anomaly characterized by multifocal congenital and progressive vascular lesions of the skin, gastrointestinal tract, and occasionally other anatomic sites, causing potentially life-threatening thrombocytopenic coagulopathy. Macroscopically, the lesions appear as round to oval, red-brown plaques, as large as a few centimeters in diameter. Histopathologically, they consist of dilated, thin-walled vessels with variable endothelial hyperplasia, positive for lymphatic endothelial cell markers, and resembling benign lymphangioendothelioma.

Dane

Klasyfikacja

Choroba

Synonimy

Cutaneovisceral angiomatosis-
thrombocytopenia syndrome

Limfangioendoteliomatoza wielogniskowa z
małopłytkowością

MLT

Zespół naczynekowości trzewno-skórnej i
małopłytkowości

MLT

Multifocal lymphangioendotheliomatosis with
thrombocytopenia

Kod ORPHA

464321

Kod OMIM

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Kod ICD10

D18.1

Kod ICD11

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*Źródło

orphanet