Opis choroby *

Definicja

A rare genetic primary lymphedema characterized by unilateral or bilateral lower limb lymphedema of variable severity. The condition shows almost complete penetrance with onset in childhood or adolescence in females, whereas in males it shows incomplete penetrance with later onset of disease. Lymphoscintigraphy in more severely affected individuals reveals lymphatic abnormalities consistent with lymphangiectasia, valve dysfunction, and thoracic duct reflux.

Dane

Klasyfikacja
Choroba

Kod ORPHA
569816

Kod OMIM
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Kod ICD10
Q82.0

Kod ICD11
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*Źródło

orphanet