

## Opis choroby \*

### Definicja

A rare genetic coagulation disorder characterized by marked bleeding tendency and posttraumatic bleeding with easy bruising, soft tissue and muscle bleeding, hemarthroses, and menorrhagia due to an increase of soluble thrombomodulin in plasma with subsequent protein C activation and reduction of thrombin generation within a potential thrombus. Abnormal laboratory findings include markedly elevated plasma thrombomodulin, reduced prothrombin consumption, and decreased thrombin generation.

### Dane

#### Klasyfikacja

Choroba

#### Synonimy

THBD-related bleeding disorder  
Koagulopatia zależna od THBD  
Koagulopatia zależna od trombomoduliny  
Zaburzenia krzepnięcia zależne od THBD  
THBD-related coagulopathy  
Thrombomodulin-related coagulopathy

#### Kod ORPHA

436169

#### Kod OMIM

614486

#### Kod ICD10

D68.3

#### Kod ICD11

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#### \*Źródło

orphanet