

## Opis choroby \*

### Definicja

A rare hereditary angioedema characterized by potentially life-threatening episodes of subcutaneous and/or submucosal edema without urticaria and with normal levels and function of C1 esterase inhibitor. Patients present with prolonged attacks which last for approximately two to five days and may include nonpitting edema of the skin, severe abdominal symptoms such as pain and swelling, and/or respiratory distress due to upper respiratory airways involvement. Affected locations and frequency of attacks differ slightly between subtypes. Estrogen-containing oral contraceptives and pregnancy are precipitating factors, especially in patients with a factor XII mutation.

### Dane

#### Klasyfikacja

Choroba

#### Synonimy

HAE with normal C1 inhibitor  
HAE z prawidłowym poziomem C1Inh  
HAE z prawidłowym poziomem inhibitora C1  
Dziedziczny obrzęk naczynioruchowy z prawidłowym poziomem inhibitora C1  
Dziedziczny obrzęk angioneurotyczny z prawidłowym poziomem C1Inh  
Dziedziczny obrzęk angioneurotyczny z prawidłowym poziomem inhibitora C1  
HAE with normal C1Inh  
Hereditary angioedema with normal C1 inhibitor  
Hereditary angioneurotic edema with normal C1 inhibitor  
Hereditary angioneurotic edema with normal C1Inh

#### Kod ORPHA

528647

#### Kod OMIM

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#### Kod ICD10

T78.3

#### Kod ICD11

4A00.14

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