

## Opis choroby \*

### Definicja

A form of junctional epidermolysis bullosa characterized by onset in childhood or young adulthood of blistering that first occurs around nails, accompanied by nail dystrophy and shedding, and then affects the hands and feet and, to a lesser extent, the elbows, and knees. Lesions heal with atrophic scarring. Other manifestations include disappearance of dermatoglyphs and palmoplantar hyperhidrosis. Extracutaneous involvement is restricted to soft tissue abnormalities of the oral cavity and enamel defects with development of caries.

### Dane

#### Klasyfikacja

Choroba

#### Synonimy

Epidermolysis bullosa progressiva

EB postępujące

JEB-lo

JEB-lo

Late-onset JEB

#### Kod ORPHA

79406

#### Kod OMIM

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#### Kod ICD10

Q81.8

#### Kod ICD11

EC31

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#### \*Źródło

orphanet