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