Opis choroby *

Definicja

A group of inherited epidermolysis bullosa (EB) characterized by cutaneous and mucosal fragility resulting in blisters and superficial ulcerations that develop below the lamina densa of the cutaneous basement membrane and that heal with significant scarring and milia formation. Dystrophic epidermolysis bullosa (DEB) comprises four major and several rare sub-types with the three most common being intermediate dominant DEB, severe recessive DEB and intermediate recessive DEB.

Dane

Klasyfikacja
Grupa fenomenów

Synonimy
DEB
DEB
Dermolytic epidermolysis bullosa
Epidermolysis bullosa dystrophica

Kod ORPHA
303

Kod OMIM
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Kod ICD10
Q81.2

Kod ICD11
EC32

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*Źródło

orphanet