Opis choroby *

Definicja

A rare, chronic, incurable, sub epithelial autoimmune bullous disease characterized by the presence of tissue bound autoantibodies against type VII collagen within the basement membrane zone of the dermal-epidermal junction of stratified squamous epithelia. The patient's serum may also have anti-type VII collagen autoantibodies. The clinical presentation is varied, and may involve the skin, oral mucosa and the upper third of the esophagus. The classical presentation is reminiscent of hereditary dystrophic epidermolysis bullosa (EB) with skin fragility, blisters and erosions and skin scarring. Other non-classical clinical presentations include an inflammatory bullous pemphigoid-like eruption, a mucous membrane pemphigoid-like eruption, and an IgA bullous dermatosis-like disease.

Dane

Klasyfikacja
Choroba

Synonimy
Acquired epidermolysis bullosa
Epidermolysis bullosa acquisita

Kod ORPHA
46487

Kod OMIM
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Kod ICD10
L12.3

Kod ICD11
EB43

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*Źródło

orphanet