

Title: Dystrophic Epidermolysis Bullosa *GeneReview* – EBS and JEB Nomenclature

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Note: The following information is provided by the authors and has not been reviewed by *GeneReviews* staff.

Table 3 – Supplementary. Comparison of 2008 Nomenclature with Proposed "Onion Skin" Terminology – Representative Examples for Epidermolysis Bullosa Simplex (EBS) and Junctional Epidermolysis Bullosa (JEB)

Old Name (per 2008 recommendations)	2014 Nomenclature
EBS, localized	EBS localized, normal keratin 5 and 14 staining, <i>KRT5</i> or <i>KRT14</i> mutation (specify type)
EBS, Dowling-Meara	EBS generalized severe, normal keratin 5 and 14 staining, <i>KRT5</i> or <i>KRT14</i> mutation (specify type)
EBS, generalized other	EBS generalized intermediate, normal keratin 5 and 14 staining, <i>KRT5</i> or <i>KRT14</i> mutation (specify type)
EBS-MP	EBS-MP, normal keratin 5 staining, <i>KRT5</i> mutation (specify type)
JEB, Herlitz	JEB generalized severe, laminin-332 absent, <i>LAMA3</i> , <i>LAMB3</i> , or <i>LAMC2</i> mutations (specify type)
JEB, non-Herlitz	JEB generalized intermediate, laminin-332 or collagen XVII reduced staining, <i>LAMA3</i> , <i>LAMB3</i> , <i>LAMC2</i> , or <i>COL17A1</i> mutations (specify type)

EBS = epidermolysis bullosa simplex

JEB = junctional epidermolysis bullosa

MP = mottled pigmentation