DEFINITION

Epidermolysis bullosa (EB) encompasses a group of otherwise heterogeneous diseases characterised by the development of blisters after minor mechanical trauma to the skin. It is the prototypic mechanobullous disease. Most cases are inherited; rare cases are acquired.

The inherited form of EB has three major subtypes-EB simplex (EBS), junctional EB (JEB), and dystrophic EB (DEB)-based on whether blisters arise in the epidermis or in the middle (i.e., the lamina lucida) or undersurface of the lowermost portion (i.e., the lamina densa) of the dermoepidermal junction, respectively. By currently accepted convention, individual subtypes of inherited EB are further defined by their model of inheritance, ultrastructural site of skin cleavage, appearance and number of specific structures in keratinocytes and the adjacent dermoepidermal junction, and the presence or absence of specific cutaneous and extracutaneous manifestations.

A noninherited form of EB, referred to as EB acquisita, is defined as a subepidermal bullous dermatosis characterised by autoimmunity to type VII collagen, the major structural component of the anchoring fibril.