



Epidermolysis Bullosa

On the basis of clinical, histologic, and electron microscopic findings, three groups of EB are recognized

In all types of EB, the blisters form as a result of minor trauma. Because of the great differences in prog

or generalized *EB dystrophica-recessiva*, a prenatal biopsy at 18 to 20 weeks of gestation is rec

There is more variability in the clinical course in some forms of EB than in others. The most severe form is

Histopathology .

□ If a fresh blister is available, a specimen for biopsy may be taken from its edge.

Even though electron microscopic examination (discussed later) is informative, the light microscopic features

In *epidermal EB*, which includes EB simplex, EB offset and hands of Webber

In *junctional EB*, the trauma of having a specimen taken for biopsy gener

the PAS-positive basement membrane zone usually remaining with the dermis . In some cases of EB let

In *EB dystrophica-dominant* and *EB dystrophica-recessive* light mic

membrane zone often appears hazy . If recognizable, it is seen in contact with the detached epidermis o

EB acquisita is not a genodermatosis but an autoimmune disorder .

Pathogenesis. If possible, all specimens of artificially induced blisters should be subjected to el

In the epidermal types of EB, electron microscopic examination shows that cleavage is the result of deg

nucleus (EM 4). Immunofluorescence mapping shows that all three antigens (type IV collagen, laminin, k

In the junctional types of EB, electron microscopic examination often shows the hemidesmosomes to be

abnormalities of the hemidesmosomes are a secondary phenomenon and the basic cause of the up

The dermal types of EB, on electron microscopy, show abnormalities in regard to their anchoring fibrils.

□

