

Bullous disorders due to hereditary or acquired desmosome or hemidesmosome impairment

A short survey

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S U M M A R Y

Some aspects of the pathogenetic mechanisms of autoimmune bullous disorders as well as of bullous hereditary disorders are shortly reviewed. The known components of desmosomes and hemidesmosomes, to which specific autoantibodies are directed in autoimmune disorders, are listed. The molecular deficiencies of desmosome and hemidesmosome components incriminated to cause hereditary bullous disorders, are also mentioned. The authors believe that clinicians should be familiar with the newest development in basic sciences concerning the pathogenetic role of desmosome and hemidesmosome.

K E Y W O R D S

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Introduction

Bullous skin disorders especially pemphigus and bullous pemphigoid presented unsurpassed therapeutic problems to dermatologists until the late fifties, when corticosteroids were introduced. The prognosis became additionally more favorable by simultaneous use of corticosteroids and immunosuppressives. Numerous studies have proven that autoimmunity is the main pathogenetic mechanism in acquired bullous diseases, whereas DNA mutations are responsible in hereditary bullous disorders. Many details remain however still to be cleared.

In the current literature our readers frequently encounter information on desmosome and hemidesmosome components, which are mentioned as the main

targets or pathogenetic factors in bullous skin disorders. In order to make more transparent to our readers, which component is linked to a given bullous dermatosis, we tried to review shortly the problem using a few schemes and tables. We realize that this is a rather difficult task as only the active investigators understand these problems in details and even their opinions sometimes differ. Franke stressed it during his lecture at the 39th ESDR Annual Meeting in Berlin that the physicochemical and immunologic characteristics of an isolated component depend at least partially on the methods applied for its isolation.

Epidermis, basal membrane and associated tissues represent living systems, which are constantly under-

