Autoimmune blistering diseases represent a group of rare, acquired disorders characterized by overlapping features, resistance to treatment, and potential lethality. Relatively recent research initiatives in laboratories throughout the World have yielded notable advances in the understanding of these diseases, including their nosology, pathophysiology, associations, and epidemiology. These advances were almost exclusively based on the study of affected patients (i.e., applied translational research). More specifically, light microscopy studies showed that the pemphigus group of diseases was characterized by intraepidermal blister formation resulting from cell-cell dysadhesion, while the pemphigoid group of diseases was characterized by subepidermal blister formation resulting from cell-matrix dysadhesion. Immunofluorescence microscopy studies showing that patients with these diseases have in situ deposits of immunoreactants in skin at the site of blister formation as well as circulating autoantibodies that bind normal human skin at the same sites demonstrated that these disorders are autoimmune in nature and characterized by a specific loss of tolerance to autoantigens in skin (Figure 1). Studies showing that autoantibodies were specific for patients with a given disease allowed such immunoreactants to serve as disease “markers” as well as probes for the recovery, characterization, and cloning of corresponding target autoantigens. Interestingly, such target autoantigens typically represent important structural proteins in skin that mediate either cell-cell or cell-matrix adhesion – vital biological processes that have been shown to be directly impaired by the effect of patients’ autoantibodies. Immunobullous disorders are among the best understood autoimmune diseases in man. Basic and translational research in this field have advanced understanding of autoimmunity and fostered the application of new and advanced therapies. The following articles review this subject in detail and provide insights about the future of research in this and related content areas.

TO CITE THIS ARTICLE

Figure 1. Immunofluorescence microscopy studies of 1 M NaCl split skin allow regional mapping of autoantibody reactivity against human epidermal basement membrane. Immunofluorescence microscopy of 1 M NaCl split skin showing a site of focal separation within the epidermal basement membrane delineated by type IV collagen (stained red) and bullous pemphigoid antigen 2 (stained green) in the base and roof, respectfully, of the test substrate. Nuclei (counterstained blue) outline the overlying epidermis as well as selected cells in the dermis.