



COL17A1 Monoclonal Antibody

Catalog No	YP-mAb-17112
Isotype	IgG
Reactivity	Human;Mouse;Rat
Applications	WB
Gene Name	COL17A1 BP180 BPAG2
Immunogen	The antiserum was produced against synthesized peptide derived from the Internal region of human COL17A1. AA range:481-530
Specificity	The antibody detects endogenous COL17A1 protein
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-1:2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	ERED;JEB4;BP180;BPA-2;BPAG2;LAD-1;BA16H23.2;Collagen XVII/COL17A1; COL17A1;COL17A1 BP180 BPAG2;collagen;type XVII;alpha 1; type XVII; alpha 1
Observed Band	150kD
Calculated Molecular Weight	150kD
Cell Pathway	Cell junction, hemidesmosome. Membrane; Single-pass type II membrane protein. Localized along the plasma membrane of the hemidesmosome.; [120 kDa linear IgA disease antigen]: Secreted, extracellular space, extracellular matrix, basement membrane. Exclusively localized to anchoring filaments. Localized to the epidermal side of split skin.; [97 kDa linear IgA disease antigen]: Secreted, extracellular space, extracellular matrix, basement membrane. Localized in the lamina lucida beneath the hemidesmosomes.
Tissue Specificity	Detected in skin (PubMed:8618013). In the cornea, it is detected in the epithelial basement membrane, the epithelial cells, and at a lower level in stromal cells (at protein level) (PubMed:25676728). Stratified squamous epithelia. Found in hemidesmosomes. Expressed in cornea, oral mucosa, esophagus, intestine, kidney collecting ducts, ureter, bladder, urethra and thymus but is absent in lung, blood vessels, skeletal muscle and nerves.



Function

disease: Defects in COL17A1 are a cause of generalized atrophic benign epidermolysis bullosa (GABEB) [MIM:226650]. GABEB is a non-lethal, adult form of junctional epidermolysis bullosa characterized by life-long blistering of the skin, associated with hair and tooth abnormalities. function: May play a role in the integrity of hemidesmosome and the attachment of basal keratinocytes to the underlying basement membrane. function: The 120 kDa linear IgA disease antigen is an anchoring filament component involved in dermal-epidermal cohesion. Is the target of linear IgA bullous dermatosis autoantibodies. miscellaneous: Both the 120 kDa linear IgA disease antigen and the 97 kDa linear IgA disease antigen of COL17A1, represent major antigenic targets of autoantibodies in patients with linear IgA disease (LAD). LAD is a subepidermal blistering disorder characterized by tissue-bound and circulating I

Background

This gene encodes the alpha chain of type XVII collagen. Unlike most collagens, collagen XVII is a transmembrane protein. Collagen XVII is a structural component of hemidesmosomes, multiprotein complexes at the dermal-epidermal basement membrane zone that mediate adhesion of keratinocytes to the underlying membrane. Mutations in this gene are associated with both generalized atrophic benign and junctional epidermolysis bullosa. Two homotrimeric forms of type XVII collagen exist. The full length form is the transmembrane protein. A soluble form, referred to as either ectodomain or LAD-1, is generated by proteolytic processing of the full length form. [provided by RefSeq, Jul 2008],

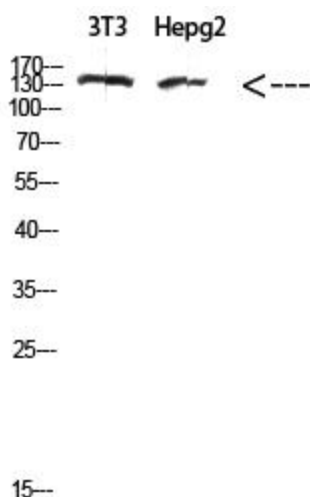
matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western Blot analysis of various cells using COL17A1 Monoclonal Antibody