Product Name: **COL17A1 Rabbit Polyclonal Antibody**
Catalog #: **APRab09173**
For research use only.

Summary

Description	Rabbit polyclonal Antibody
Host	Rabbit
Application	WB,ICC/IF,ELISA
Reactivity	Human,Rat,Mouse
Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Concentration	1mg/ml
Storage	Aliquot and store at -20°C (valid for 12 months). Avoid freeze/thaw cycles.
Shipping	Ice bags
Buffer	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
Purification	Affinity purification

Application

Dilution Ratio	WB 1:500-1:2000,ICC/IF 1:100-1:300,ELISA 1:5000-1:20000
Molecular Weight	150kDa

Antigen Information

Gene Name	COL17A1 BP180 BPAG2
Alternative Names	COL17A1 BP180 BPAG2
Gene ID	1308.0
SwissProt ID	Q9UMD9
Immunogen	The antiserum was produced against synthesized peptide derived from the Internal region of human COL17A1. AA range:481-530

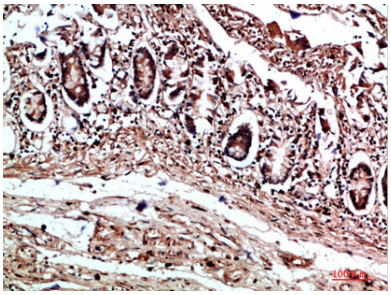
Background

This gene encodes the alpha chain of type XVII collagen. Unlike most collagens, collagen XVII is a transmembrane protein.

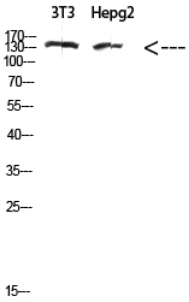
Collagen XVII is a structural component of hemidesmosomes, multiprotein complexes at the dermal-epidermal basement membrane zone that mediate adhesion of keratinocytes to the underlying membrane. Mutations in this gene are associated with both generalized atrophic benign and junctional epidermolysis 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],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 IgA autoantibodies to the dermal-epidermal junction. These IgA autoantibodies preferentially react with 97 and the 120 kDa forms, but not with the full-length COL17A1, suggesting that the cleavage of the ectodomain generates novel autoantigenic epitopes.,PTM:Prolines at the third position of the tripeptide repeating unit (G-X-Y) are hydroxylated in some or all of the chains.,PTM:The ectodomain is shedded from the surface of keratinocytes resulting in a 120-kDa soluble form, also named as 120 kDa linear IgA disease antigen. The shedding is mediated by membrane-bound metalloproteases. This cleavage is inhibited by phosophorylation at Ser-544.,PTM:The intracellular/endo domain is disulfide-linked.,sequence caution:Contaminating sequence. Potential poly-A sequence.,subcellular location:Exclusively localized to anchoring filaments. Localized to the epidermal side of split skin.,subcellular location:Localized along the plasma membrane of the hemidesmosome.,subcellular location:Localized in the lamina lucida beneath the hemidesmosomes.,subunit:Homotrimers of alpha 1(XVII)chains.,tissue specificity: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.,

Research Area

Image Data



Immunohistochemical analysis of paraffin-embedded human-colon, antibody was diluted at 1:200



Western Blot analysis of 3T3, hepg2 cells using Antibody diluted at 500. Secondary antibody was diluted at 1:20000