

产品名称: COL17A1 Rabbit Polyclonal Antibody
产品货号: APRab09173



产品概述 (Summary)

产品名称 (Production Name)	COL17A1 Rabbit Polyclonal Antibody
描述 (Description)	Rabbit polyclonal Antibody
宿主 (Host)	Rabbit
应用 (Application)	WB, ICC/IF, ELISA
种属反应性 (Reactivity)	Human, Rat, Mouse

产品性能 (Performance)

偶联物 (Conjugation)	Unconjugated
修饰 (Modification)	Unmodified
同种型 (Isotype)	IgG
克隆 (Clonality)	Polyclonal
形式 (Form)	Liquid
存放说明 (Storage)	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
储存溶液 (Buffer)	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
纯化方式 (Purification)	Affinity purification

免疫原信息 (Immunogen)

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

产品应用 (Application)

稀释比 (Dilution Ratio)	WB 1:500-1:2000, ICC/IF 1:100-1:300, ELISA 1:5000-1:20000
蛋白分子量 (Molecular Weight)	150kDa

研究背景 (Background)

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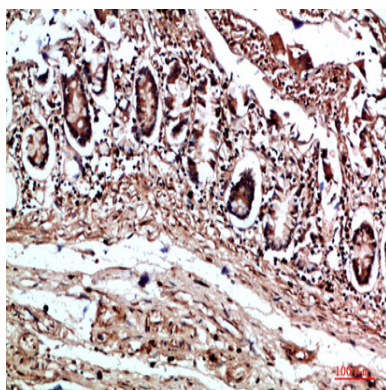


This gene encodes the alpha chain of type XVII collagen. Unlike most collagens, collagen XVII is a transmembrane protein. Collagen XVII is a structural component of hemidesmosomes, multiprotein complexes at the dermal-epidermal basement membrane zone that mediate adhesion of keratinocytes to the underlying 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 phosphorylation 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.,

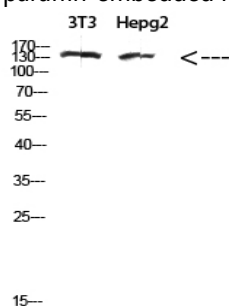
研究领域 (Research Area)

图片 (Image Data)

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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

注意事项 (Note)

For research use only .