产品名称：COL7A1 Rabbit Polyclonal Antibody
产品货号：APRab09198

产品概述（Summary）

产品名称（Production Name）	COL7A1 Rabbit Polyclonal Antibody
描述（Description）	Rabbit polyclonal Antibody
宿主（Host）	Rabbit
应用（Application）	IHC,ICC/IF,ELISA
种属反应性（Reactivity）	Human,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）	COL7A1
别名（Alternative Names）	COL7A1; Collagen alpha-1(VII) chain; Long-chain collagen; LC collagen
基因 ID（Gene ID）	1294.0
蛋白 ID（SwissProt ID）	Q02388.The antiserum was produced against synthesized peptide derived from human Collagen VII alpha1. AA range:1841-1890

产品应用（Application）

稀释比（Dilution Ratio）	IHC 1:100-1:300,ICC/IF 1:200-1:1000,ELISA 1:5000-1:10000
蛋白分子量（Molecular Weight）	

研究背景（Background）

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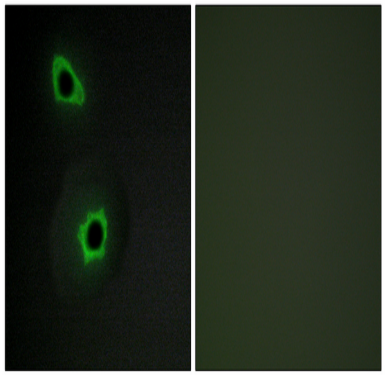
collagen type VII alpha 1 chain(COL7A1) Homo sapiens　This gene encodes the alpha chain of type VII collagen. The type VII collagen fibril, composed of three identical alpha collagen chains, is restricted to the basement zone beneath stratified squamous epithelia. It functions as an anchoring fibril between the external epithelia and the underlying stroma. Mutations in this gene are associated with all forms of dystrophic epidermolysis bullosa. In the absence of mutations, however, an acquired form of this disease can result from an autoimmune response made to type VII collagen. [provided by RefSeq, Jul 2008],disease:Defects in COL7A1 are the cause of epidermolysis bullosa dystrophica (DEB) [MIM:131750, 226600]. DEB defines a group of blistering skin diseases characterized by tissue separation which occurs below the dermal-epidermal basement membrane at the level of the anchoring fibrils. Inheritance can be autosomal dominant or recessive. Various clinical types with different severity are recognized, ranging from severe mutilating forms to mild forms with limited and localized scarring, and less frequent extracutaneous manifestations. Mild forms include epidermolysis bullosa mitis and epidermolysis bullosa localisata.,disease:Defects in COL7A1 are the cause of epidermolysis bullosa dystrophica Bart type (B-DEB) [MIM:132000]. B-DEB is an autosomal dominant form of dystrophic epidermolysis bullosa characterized by congenital localized absence of skin, skin fragility and deformity of nails.,disease:Defects in COL7A1 are the cause of epidermolysis bullosa dystrophica Hallopeau-Siemens type (HS-DEB) [MIM:226600]. HS-DEB is the most severe recessive form and manifests with mutilating scarring, joint contractures, corneal erosions, esophagus structures, and propensity to formation of cutaneous squamous cell carcinomas leading to premature demise of the affected individuals.,disease:Defects in COL7A1 are the cause of epidermolysis bullosa dystrophica Pasini type (P-DEB) [MIM:131750]; also known as albopapuloid dominant dystrophic epidermolysis bullosa. P-DEB is a severe, dominantly inherited form of dystrophic epidermolysis bullosa characterized by albopapuloid Pasini papule, dorsal extremity blistering, milia formation and red atrophic scarring after recurrent blisters.,disease:Defects in COL7A1 are the cause of epidermolysis bullosa dystrophica pretibial type (PR-DEB) [MIM:131850]. PR-DEB is characterized by pretibial blisters that develop into prurigo-like hyperkeratotic lesions. It predominantly affects the pretibial areas, sparing the knees and other parts of the skin. Other clinical features include nail dystrophy, albopapuloid skin lesions, and hypertrophic scars without pretibial predominance. The phenotype shows considerable interindividual variability Inheritance is autosomal dominant.,disease:Defects in COL7A1 are the cause of epidermolysis bullosa dystrophica with subcorneal cleavage (EBDSC) [MIM:607600]; also known as epidermolysis bullosa simplex superficialis (EBSS). EBDSC is a new variant of epidermolysis bullosa simplex (EBS), characterized by the development of skin cleavage just beneath the level of stratum corneum. It appears to be transmitted as an autosomal dominant trait and differs from other autosomal dominant forms of EBS by the common findings of milia and atrophic scarring, as well as involvement of oral and/or ocular surfaces. It is further differentiated by the presence of blisters and the absence of spontaneous continual exfoliation or peeling.,disease:Defects in COL7A1 are the cause of epidermolysis bullosa pruriginosa (EBP) [MIM:604129]. EBP is a distinct clinical subtype of DEB. It is characterized by skin fragility, blistering, scar formation, intense pruritus and excoriated prurigo nodules. Onset is in early childhood, but in some cases is delayed until the second or third decade of life. Inheritance can be autosomal dominant or recessive.,disease:Defects in COL7A1 are the cause of isolated toenail dystrophy without skin fragility [MIM:607523].,disease:Defects in COL7A1 are the cause of transient bullous dermolysis of the newborn (TBDN) [MIM:131705]. TBDN is a neonatal form of dystrophic epidermolysis bullosa characterized by sub-epidermal blisters, reduced or abnormal anchoring fibrils at the dermo-epidermal junction,

and electron-dense inclusions in keratinocytes. TBDN heals spontaneously or strongly improves within the first months and years of life.,disease:Epidermolysis bullosa acquisita (EBA) is an autoimmune acquired blistering skin disease resulting from autoantibodies to type VII collagen.,function:Stratified squamous epithelial basement membrane protein that forms anchoring fibrils which may contribute to epithelial basement membrane organization and adherence by interacting with extracellular matrix (ECM) proteins such as type IV collagen.,PTM:Prolines at the third position of the tripeptide repeating unit (G-X-Y) are hydroxylated in some or all of the chains.,similarity:Contains 1 BPTI/Kunitz inhibitor domain.,similarity:Contains 2 VWFA domains.,similarity:Contains 9 fibronectin type-III domains.,subunit:Homotrimer. Interacts with MIA3/TANGO1; facilitating its loading into transport carriers and subsequent secretion.,

研究领域（Research Area）

图片（Image Data）



Immunofluorescence analysis of COS7 cells, using Collagen VII alpha1 Antibody. The picture on the right is blocked with the synthesized peptide.

注意事项（Note）

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