

产品名称: COL6A3 Rabbit Polyclonal Antibody

产品货号: APRab09197

产品概述 (Summary)

描述(Description)	Rabbit polyclonal Antibody
宿主(Host)	Rabbit
应用(Application)	IHC, ICC/IF, ELISA
种属反应(Reactivity)	Human, Mouse
偶联物(Conjugation)	Unconjugated
修饰(Modification)	Unmodified
同种型(Isotype)	IgG
克隆(Clonality)	Polyclonal
剂型(Form)	Liquid
保存条件(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 (Purification)	Affinity purification

产品应用(Application)

稀释比(Dilution Ratio) IHC 1:100-1:300, ICC/IF 1:200-1:1000, ELISA 1:20000-1:40000

分子量(Molecular Weight)

抗原信息(Antigen Information)

基因名(Gene Name)	COL6A3
别名(Alternative Names)	COL6A3; Collagen alpha-3(VI) chain
基因 ID(Gene ID)	1293.0
SwissProt ID	P12111
免疫原(Immunogen)	The antiserum was produced against synthesized peptide derived from human Collagen VI alpha3. AA range:2261-2310

研究背景 (Background)

This gene encodes the alpha-3 chain, one of the three alpha chains of type VI collagen, a beaded filament collagen found in most connective tissues. The alpha-3 chain of type VI collagen is much larger than the alpha-1 and -2 chains. This difference in

size is largely due to an increase in the number of subdomains, similar to von Willebrand Factor type A domains, that are found in the amino terminal globular domain of all the alpha chains. These domains have been shown to bind extracellular matrix proteins, an interaction that explains the importance of this collagen in organizing matrix components. Mutations in the type VI collagen genes are associated with Bethlem myopathy, a rare autosomal dominant proximal myopathy with early childhood onset. Mutations in this gene are also a cause of Ullrich congenital muscular dystrophy, also referred to as Ullrich scleroatonic muscular dystrophy, an adisease:Defects in COL6A3 are a cause of Bethlem myopathy (BM) [MIM:158810]. BM is a rare autosomal dominant proximal myopathy characterized by early childhood onset (complete penetrance by the age of 5) and joint contractures most frequently affecting the elbows and ankles.,disease:Defects in COL6A3 are a cause of Ullrich congenital muscular dystrophy (UCMD) [MIM:254090]; also known as Ullrich scleroatonic muscular dystrophy. UCMD is an autosomal recessive congenital myopathy characterized by muscle weakness and multiple joint contractures, generally noted at birth or early infancy. The clinical course is more severe than in Bethlem myopathy.,function:Collagen VI acts as a cell-binding protein.,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 N-terminus is blocked.,similarity:Belongs to the type VI collagen family.,similarity:Contains 1 BPTI/Kunitz inhibitor domain.,similarity:Contains 1 fibronectin type-III domain.,similarity:Contains 12 VWFA domains.,similarity:Contains 16 LRR (leucine-rich) repeats.,similarity:Contains 5 collagen-like domains.,subunit:Trimers composed of three different chains: alpha-1(VI), alpha-2(VI), and alpha-3(VI) or alpha-5(VI) or alpha-6(VI),.

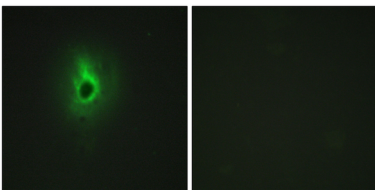
研究领域 (Research Area)

Focal adhesion;ECM-receptor interaction;

注意事项 (Note)

For research use only.

图片 (Image Data)



Immunofluorescence analysis of HeLa cells, using Collagen VI alpha3 Antibody. The picture on the right is blocked with the synthesized peptide.

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产品类别	产品货号	产品名称
WB 解决方案	RA10020	2h 极速 WB 即用型全流程试剂盒
	RA10021	4h 快速 WB 即用型全流程试剂盒
	RA10042	5h 畅享版 WB 全流程试剂盒
	RA10037	校准级彩色预染蛋白 Marker (8-180kDa)
	RA10038	校准级彩色预染蛋白 Marker (10-250kDa)
	RA10039	校准级高分子彩色预染蛋白 Marker (25-400kDa)
TSA 多重荧光染色试剂盒	RA10008	TSA 双标三色多重荧光染色试剂盒 (mIHC)
	RA10009	TSA 三标四色多重荧光染色试剂盒 (mIHC)
	RA10010	TSA 四标五色多重荧光染色试剂盒 (mIHC)
	RA10011	TSA 五标六色多重荧光染色试剂盒 (mIHC)
	RA10012	TSA 六标七色多重荧光染色试剂盒 (mIHC)
IHC 检测试剂盒	RA10006	HRP Anti-Mouse/Rabbit IHC Detection System
	RA10007	Polymer-HRP Anti-Mouse/Rabbit IHC Detection System
抗体标记试剂盒	RE80004p	辣根过氧化物酶(HRP)抗体标记试剂盒
	RE80002q	Sulfo-NHS-生物素标记试剂盒
	RE80007p	Cy3 荧光素标记试剂盒
	RE80011p	Fluor488 荧光素标记试剂盒
	RE80017p	Fluor750 荧光素标记试剂盒
	RE80005p	藻红蛋白(R-PE) 抗体快速标记试剂盒
	RE80040	PE-Cy7 串联染料抗体快速标记试剂盒
稳转细胞系构建服务 (免费赠送全膜 WB 验证)	TS-0001	过表达稳转细胞系构建
	TS-0002	敲低稳转细胞系构建
	TS-0003	敲除细胞系构建

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