**产品名称：COL6A3 Rabbit Polyclonal Antibody**
**产品货号：APRab09197**

## 产品概述（Summary）

| | |
|---|---|
| **产品名称（Production Name）** | COL6A3 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）

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

## 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 |
| **蛋白分子量（Molecular Weight）** | |

## 研究背景（Background）

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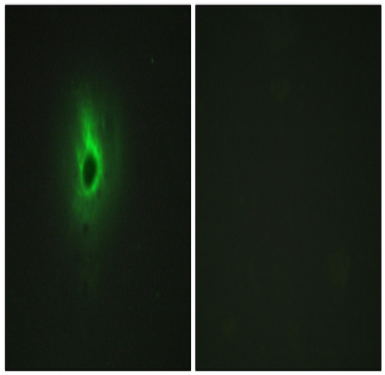
This gene encodes the alpha-3 chain, one of the three alpha chains of type VI collagen, a beaded filament collagen found in most connective tissues. The alpha-3 chain of type VI collagen is much larger than the alpha-1 and -2 chains. This difference in size is largely due to an increase in the number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

## 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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## 注意事项（Note）

For research use only .