Product Name: COL6A3 Rabbit Polyclonal Antibody
Catalog #: APRab09197

Summary

Production Name	COL6A3 Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	IF,ELISA
Reactivity	Human,Mouse

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

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

Application

Dilution Ratio	IF 1:200-1:1000. ELISA: 1:40000.
Molecular Weight	

Background

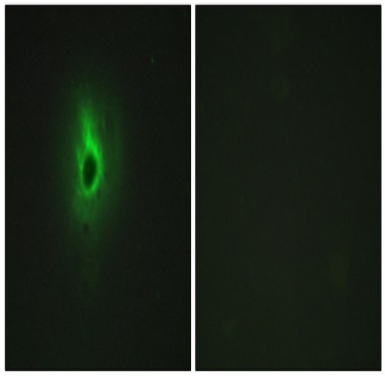
This gene encodes the alpha-3 chain, one of the three alpha chains of type VI collagen, a beaded filament collagen found

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).,

Research Area

Focal adhesion;ECM-receptor interaction;

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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Note

For research use only.