



Rabbit Anti-Collagen VI alpha 1 antibody

SL0553R

Product Name:	Collagen VI alpha 1
Chinese Name:	抗VI型胶原抗体
Alias:	CO6A1_HUMAN; COL6A1; COL6A2; COL6A3; Collagen alpha 1(VI) chain; Collagen alpha 1(VI) chain precursor; Collagen alpha-1(VI) chain; Collagen type VI alpha 1; Collagen type VI alpha 2; Collagen type VI alpha 3; Collagen VI alpha 1 polypeptide; Collagen VI alpha 2 polypeptide; Collagen VI alpha 3 polypeptide; CollagenVI; Human mRNA for collagen VI alpha 1 C terminal globular domain; OPLL; PP3610.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Rabbit,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	106kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Collagen VI:951-1028/1028
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	The collagens are a superfamily of proteins that play a role in maintaining the integrity of various tissues. Collagens are extracellular matrix proteins and have a triple-helical

domain as their common structural element. Collagen VI is a major structural component of microfibrils. The basic structural unit of collagen VI is a heterotrimer of the alpha1(VI), alpha2(VI), and alpha3(VI) chains. The alpha2(VI) and alpha3(VI) chains are encoded by the COL6A2 and COL6A3 genes, respectively. The protein encoded by this gene is the alpha 1 subunit of type VI collagen (alpha1(VI) chain). Mutations in the genes that code for the collagen VI subunits result in the autosomal dominant disorder, Bethlem myopathy. [provided by RefSeq, Jul 2008]

Function:

Collagen VI acts as a cell-binding protein.

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).

Subcellular Location:

Secreted, extracellular space, extracellular matrix (By similarity).

Tissue Specificity:

Prolines at the third position of the tripeptide repeating unit (G-X-Y) are hydroxylated in some or all of the chains.

DISEASE:

Bethlem myopathy (BM) [MIM:158810]: A benign autosomal dominant proximal myopathy characterized by early childhood onset and joint contractures most frequently affecting the elbows and ankles. Note=The disease is caused by mutations affecting the gene represented in this entry.

Ullrich congenital muscular dystrophy (UCMD) [MIM:254090]: UCMD is a 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. Note=The disease is caused by mutations affecting the gene represented in this entry.

Similarity:

Belongs to the type VI collagen family.
Contains 3 VWFA domains.

SWISS:

P12109

Gene ID:

1291

Database links:

[Entrez Gene: 1291](#) Human

[Entrez Gene: 1292](#)Human

[Entrez Gene: 1293](#)Human

[Entrez Gene: 12833](#)Mouse

[Entrez Gene: 12834](#)Mouse

[Entrez Gene: 12835](#)Mouse

[Entrez Gene: 294337](#)Rat

[Entrez Gene: 361821](#)Rat

[Omin: 120220](#)Human

[Omin: 120240](#)Human

[Omin: 120250](#)Human

[SwissProt: P12109](#)Human

[SwissProt: P12110](#)Human

[SwissProt: P12111](#)Human

[SwissProt: Q02788](#)Mouse

[SwissProt: Q04857](#)Mouse

[Unigene: 474053](#)Human

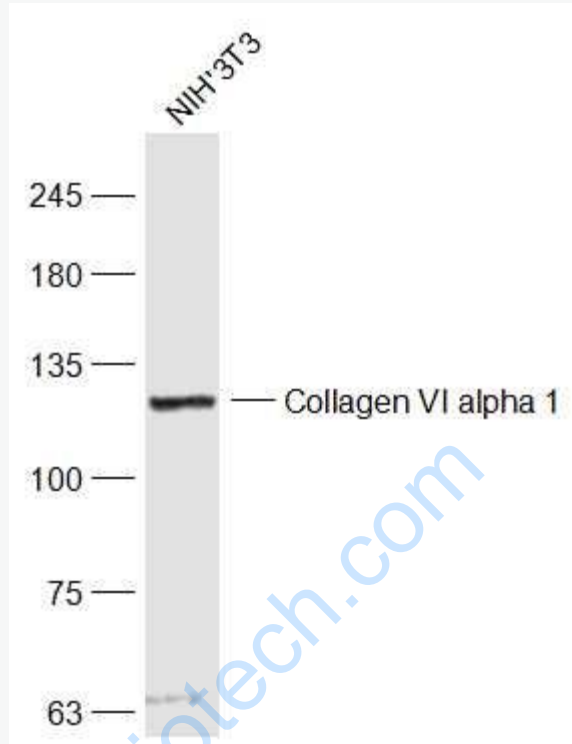
[Unigene: 2509](#)Mouse

[Unigene: 232118](#)Rat

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Picture:



Sample:

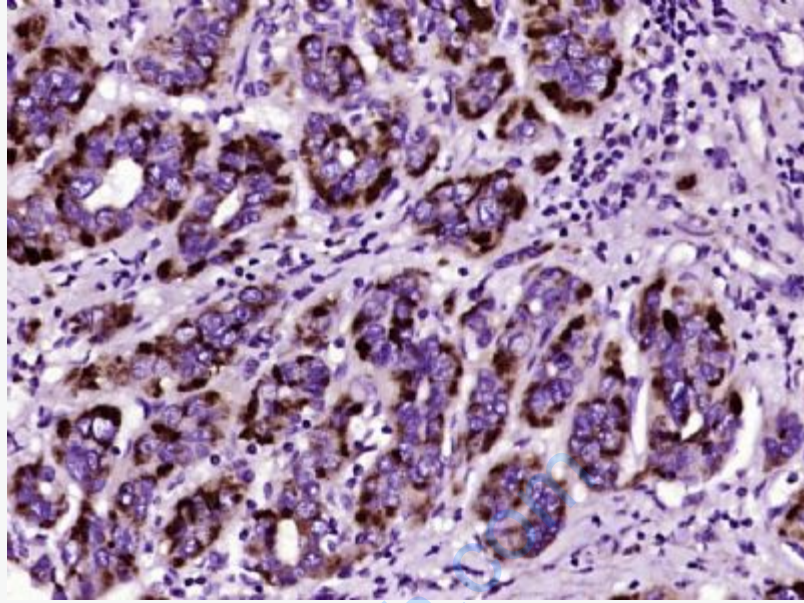
NIH/3T3(Mouse) Cell Lysate at 30 ug

Primary: Anti-Collagen VI alpha 1 (SL0553R) at 1/1000 dilution

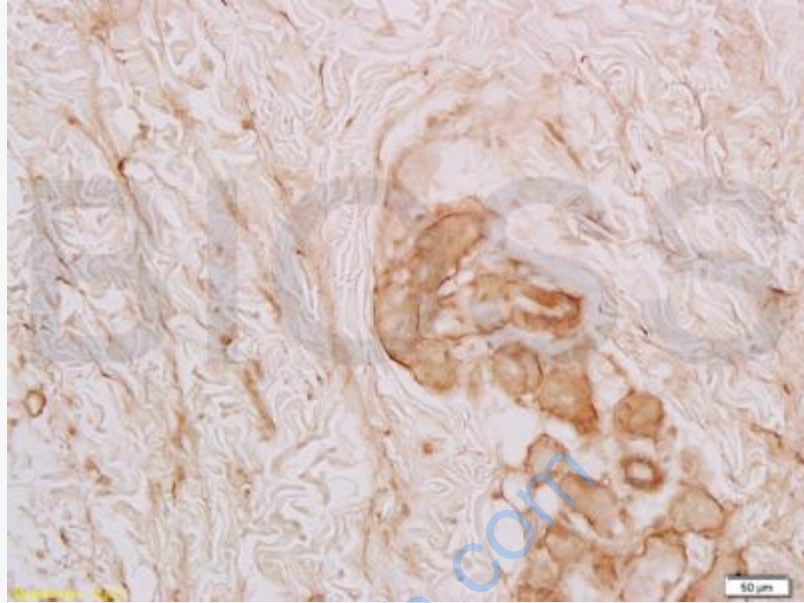
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 106 kD

Observed band size: 131 kD



Paraformaldehyde-fixed, paraffin embedded (human liver carcinoma); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Collagen VI alpha 1) Polyclonal Antibody, Unconjugated (SL0553R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.



Tissue/cell: The skin of the infant; 4% Paraformaldehyde-fixed and paraffin-embedded;

Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37°C for 20 min;

Incubation: Anti-Collagen-VI-alpha1 Polyclonal Antibody, Unconjugated(SL0553R) 1:100, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining