abcam

Product datasheet

Recombinant Human Collagen VI alpha 2 protein ab169887

Description

Product name	Recombinant Human Collagen VI alpha 2 protein
Purity	> 90 % SDS-PAGE. The final product was refolded using a unique “temperature shift inclusion body refolding” technology and chromatographically purified.
Expression system	Escherichia coli
Accession	P12110
Protein length	Protein fragment
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MASMTGGQQMGRGHHHHHHGNLYFQGGETELSVAQCTQ RPVDIVFLLDGS ERLGEQNFHKARRFVEQVARRLTLARRDDDPLNARVALL QFGGPGEQQVA FPLSHNLTAIHEALETTQYLNSFSHVGAGVVHAINAIVRSPR GRARRHAE LSFVFLTDGVTGNDSLHESAHSMRKQNVVPTVLALGSDV DMDVLTTLSLG DRAAVFHEKDYDSLAQPGFFDRFIRWIC
Predicted molecular weight	25 kDa including tags
Amino acids	820 to 1019
Tags	His-T7 tag N-Terminus

Specifications

Our Abpromise guarantee covers the use of **ab169887** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	SDS-PAGE
Form	Liquid

Preparation and Storage

Stability and Storage	Shipped at 4°C. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituent: 0.32% Tris HCl Contains NaCl, KCl, EDTA, arginine, DTT and Glycerol.

General Info

Function	Collagen VI acts as a cell-binding protein.
Involvement in disease	Defects in COL6A2 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. Defects in COL6A2 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. Defects in COL6A2 are the cause of myosclerosis autosomal recessive (MYOSAR) [MIM:255600]; also known as myosclerotic myopathy or congenital myosclerosis of Lowenthal. A condition characterized by chronic inflammation of skeletal muscle with hyperplasia of the interstitial connective tissue. The clinical picture includes slender muscles with firm 'woody' consistency and restriction of movement of many joints because of muscle contractures.
Sequence similarities	Belongs to the type VI collagen family. Contains 3 VWFA domains.
Post-translational modifications	Prolines at the third position of the tripeptide repeating unit (G-X-Y) are hydroxylated in some or all of the chains.
Cellular localization	Secreted > extracellular space > extracellular matrix. Membrane. Recruited on membranes by CSPG4.

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