abcam

Product datasheet

Native Human Collagen VI protein ab7538

4 References

Description

Product name	Native Human Collagen VI protein
Purity	> 95 % n/a. This product has been prepared from human placenta and is chromatographically and immunologically pure.
Expression system	Native
Accession	P12109
Protein length	Full length protein
Animal free	No
Nature	Native
Species	Human
Additional sequence information	Prepared from Human Placenta.

Specifications

Our Abpromise guarantee covers the use of **ab7538** in the following tested applications.

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

Applications	Western blot Immunoprecipitation ELISA Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections)
Form	Liquid
Additional notes	This product is free from other collagens, human serum proteins and non-collagen extracellular matrix proteins. This product reacts with anti-Collagen Type VI. Reaction with anti-Collagen I, II, III, IV or V is negligible (typically less than 1% cross reactivity was detected by ELISA).

Preparation and Storage

Stability and Storage	Shipped at 4°C. Store at +4°C. Diluted stock solutions should be used immediately and not be frozen. Please see notes section. Store undiluted. Preservative: 0.01% Sodium azide Constituent: 3% Acetic acid

General Info

Function	Collagen VI acts as a cell-binding protein.
Involvement in disease	Defects in COL6A1 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 COL6A1 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.
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.