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 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

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General Info

modifications

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 similaritiesBelongs to the type VI collagen family.

Contains 3 VWFA domains.

Post-translational 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.

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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