abcam

Product datasheet

Native Human Collagen VI protein ab7538

4 References

製品の詳細

製品名 Native Human Collagen VI protein

発現系Nativeアクセッション番号P12109

タンパク質長 Full length protein

Animal free No

由来 Native

生物種 Human

配列の追加情報 Prepared from Human Placenta.

特性

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.

アプリケーション SDS-PAGE

Western blot

Immunoprecipitation

ELISA

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections)

製品の状態 Liquid

備考 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).

前処理および保存

保存方法および安定性 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

関連情報

機能 Collagen VI acts as a cell-binding protein.

関連疾患 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.

配列類似性 Belongs to the type VI collagen family.

Contains 3 VWFA domains.

翻訳後修飾 Prolines at the third position of the tripeptide repeating unit (G-X-Y) are hydroxylated in some or all

of the chains.

細胞内局在 Secreted > extracellular space > extracellular matrix.

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