abcam

Product datasheet

Native Human Collagen VI protein ab7538

4 References

描述

产品名称 Native人Collagen VI蛋白

表达系统 Native Accession P12109

蛋白长度 Full length protein

无动物成分 No

性质 Native

种属 Human

额外的序列信息 Prepared from Human Placenta.

技术指标

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.

应用 SDS-PAGE

Western blot

Immunoprecipitation

ELISA

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

形式 Liquid

补充说明 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).

制备和贮存

稳定性和存储 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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常规信息

功能 Collagen VI acts as a cell-binding protein.

疾病相关 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.

序列相似性 Belongs to the type VI collagen family.

Contains 3 VWFA domains.

翻译后修饰 Prolines at the third position of the tripeptide repeating unit (G-X-Y) are hydroxylated in some or all

of the chains.

细胞定位 Secreted > extracellular space > extracellular matrix.

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

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