软骨相关蛋白 CRTAP 抗体

产品货号： mlR12949

英文名称： CRTAP

中文名称： 软骨相关蛋白 CRTAP 抗体

别　　名： Cartilage associated protein; LEPREL3; leprecan-like 3; CASP; CRTAP_HUMAN.

研究领域： 细胞生物　信号转导　细胞外基质

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应　： Human, Mouse, Rat, Chicken, Dog, Pig, Horse, Rabbit,

产品应用： WB=1:500-2000 ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500
（石蜡切片需做抗原修复）
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

分 子 量： 44kDa

细胞定位： 分泌型蛋白

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human CRTAP:201-300/401

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： Store at -20 ° C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20° C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ° C.

PubMed： PubMed

产品介绍： CRTAP is a secreted protein localizing to the extracellular space that plays a role in collagen post-translational modifications, extracellular fibril assembly and intracellular trafficking. CRTAP is widely expressed with predominant expression in articular chondrocytes. It contains a signal peptide and a tetratricopeptide-like helical domain and is essential for normal bone formation. In the endoplasmic reticulum (ER), CRTAP forms a complex with Gros1 and CyPB (cyclophilin B) and is required for the efficient 3-hydroxylation of target prolyl residues in Collagen Type I molecules, the major structural proteins of skin and bone. Mutations in the gene encoding CRTAP can lead to autosomal recessive osteogenesis imperfecta (OI) type 7 and type 2B. OI, also known as brittle bone disease, is characterized by bone fragility and susceptibility to fractures. OI type 7 is a mild form of this disorder, while OI type 2B is a neonatal lethal condition.

Function:

CRTAP is found in articular chondrocytes and is expressed in a variety of other tissues. Defects in CRTAP are the cause of osteogenesis imperfecta type 7 (OI-7). OI is a connective tissue disorder characterized by bone fragility and low bone mass. OI-7 is an autosomal recessive form of OI.

Subcellular Location:

Secreted, extracellular space, extracellular matrix (By similarity).

Tissue Specificity:

Found in articular chondrocytes. Expressed in a variety of tissues.

DISEASE:

Defects in CRTAP are the cause of osteogenesis imperfecta type 7 (OI7) [MIM:610682]. A connective tissue disorder characterized by short stature, short humeri and femora, coxa vara, white sclera, and the absence of dentinogenesis imperfecta. Multiple fractures are present at birth, and patients manifest moderate-severe bone fragility. Death may occurr in the perinatal period due to secondary respiratory insufficiency.

Similarity:

Belongs to the leprecan family.

SWISS:

O75718

Gene ID:

10491

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

产品图片