

1 型前胶原氨基末端前肽/Ⅰ型前胶原氨基末端前肽抗体

产品货号： mlR6379

英文名称： Procollagen type I N terminal propeptide

中文名称： 1 型前胶原氨基末端前肽/Ⅰ型前胶原氨基末端前肽抗体

别名： COL1A1; Alpha 1 type I collagen; Alpha-1 type I collagen; CO1A1_HUMAN; COL1A1; Collagen alpha-1(I) chain; Collagen type I alpha 1; Procollagen type I N terminal propeptide; Procollagen type I N-terminal propeptide.

研究领域： 信号转导 细胞骨架 细胞外基质

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Dog, Cow, Horse, Sheep,

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： 15kDa

细胞定位： 细胞外基质 分泌型蛋白

性状： Lyophilized or Liquid

浓 度 : 1mg/ml

免 疫 原 : KLH conjugated synthetic peptide derived from human Procollagen type I N terminal propeptide:1-100/1464

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 : This gene encodes the pro-alpha1 chains of type I collagen whose triple helix comprises two alpha1 chains and one alpha2 chain. Type I is a fibril-forming collagen found in most connective tissues and is abundant in bone, cornea, dermis and tendon. Mutations in this gene are associated with osteogenesis imperfecta types I-IV, Ehlers-Danlos syndrome type VIIA, Ehlers-Danlos syndrome Classical type, Caffey Disease and idiopathic osteoporosis. Reciprocal translocations between chromosomes 17 and 22, where this gene and the gene for platelet-derived growth factor beta are located, are associated with a particular type of skin tumor called dermatofibrosarcoma protuberans, resulting from unregulated expression of the growth factor. Two transcripts, resulting from the use of alternate polyadenylation signals, have been identified for this gene. [provided by R. Dalglish, Feb 2008].

Function:

Type I collagen is a member of group I collagen (fibrillar forming collagen).

Subunit:

Trimers of one alpha 2(I) and two alpha 1(I) chains. Interacts with MRC2. Interacts with TRAM2.

Subcellular Location:

Secreted, extracellular space, extracellular matrix.

Tissue Specificity:

Forms the fibrils of tendon, ligaments and bones. In bones the fibrils are mineralized with calcium hydroxyapatite.

Post-translational modifications:

Proline residues at the third position of the tripeptide repeating unit (G-X-P) are hydroxylated in some or all of the chains. Proline residues at the second position of the tripeptide repeating unit (G-P-X) are hydroxylated in some of the chains.

O-linked glycan consists of a Glc-Gal disaccharide bound to the oxygen atom of a post-translationally added hydroxyl group.

DISEASE:

Defects in COL1A1 are the cause of Caffey disease (CAFFD) [MIM:114000]; also known as infantile cortical hyperostosis. Caffey disease is characterized by an infantile episode of massive subperiosteal new bone formation that typically involves the diaphyses of the long bones, mandible, and clavicles. The involved bones may also appear inflamed, with painful swelling and systemic fever often accompanying the illness. The bone changes usually begin before 5 months of age and resolve before 2 years of age.

Defects in COL1A1 are a cause of Ehlers-Danlos syndrome type 1 (EDS1) [MIM:130000]; also known as Ehlers-Danlos syndrome gravis. EDS is a connective tissue disorder characterized by hyperextensible skin, atrophic cutaneous scars due to tissue fragility and joint hyperlaxity. EDS1 is the severe form of classic Ehlers-Danlos syndrome.

Defects in COL1A1 are the cause of Ehlers-Danlos syndrome type 7A (EDS7A) [MIM:130060]; also known as autosomal dominant Ehlers-Danlos syndrome type VII. EDS is a connective tissue disorder characterized by hyperextensible skin, atrophic cutaneous scars due to tissue fragility and joint hyperlaxity. EDS7A is marked by

bilateral congenital hip dislocation, hyperlaxity of the joints, and recurrent partial dislocations.

Defects in COL1A1 are a cause of osteogenesis imperfecta type 1 (OI1) [MIM:166200]. A dominantly inherited connective tissue disorder characterized by bone fragility and blue sclerae. Osteogenesis imperfecta type 1 is non-deforming with normal height or mild short stature, and no dentinogenesis imperfecta.

Defects in COL1A1 are a cause of osteogenesis imperfecta type 2 (OI2) [MIM:166210]; also known as osteogenesis imperfecta congenita. A connective tissue disorder characterized by bone fragility, with many perinatal fractures, severe bowing of long bones, undermineralization, and death in the perinatal period due to respiratory insufficiency.

Defects in COL1A1 are a cause of osteogenesis imperfecta type 3 (OI3) [MIM:259420]. A connective tissue disorder characterized by progressively deforming bones, very short stature, a triangular face, severe scoliosis, grayish sclera, and dentinogenesis imperfecta.

Defects in COL1A1 are a cause of osteogenesis imperfecta type 4 (OI4) [MIM:166220]; also known as osteogenesis imperfecta with normal sclerae. A connective tissue disorder characterized by moderately short stature, mild to moderate scoliosis, grayish or white sclera and dentinogenesis imperfecta.

Genetic variations in COL1A1 are a cause of susceptibility to osteoporosis (OSTEOP) [MIM:166710]; also known as involutional or senile osteoporosis or postmenopausal osteoporosis. Osteoporosis is characterized by reduced bone mass, disruption of bone microarchitecture without alteration in the composition of bone. Osteoporotic bones are more at risk of fracture.

Note=A chromosomal aberration involving COL1A1 is found in dermatofibrosarcoma protuberans. Translocation t(17;22)(q22;q13) with PDGF.

Similarity:

Belongs to the fibrillar collagen family.

Contains 1 fibrillar collagen NC1 domain.

Contains 1 VWFC domain.

SWISS:

P02452



Gene ID:

1277

Important Note:

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