胞外基质蛋白 3 抗体

产品货号： mlR18694

英文名称： Matrilin 3

中文名称： 胞外基质蛋白 3 抗体

别　　名： AV009181; DIPOA; EDM5; HOA; MATN3; MATN3_HUMAN; Matrilin 3; Matrilin-3; OADIP; OS2.

研究领域： 细胞生物　发育生物学　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, Dog, Pig, Cow, Horse, Rabbit, Guinea Pig, Cat,

产品应用： 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： 53kDa

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 Matrilin 3:51-150/486

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 a member of von Willebrand factor A domain containing protein family. This family of proteins is thought to be involved in the formation of filamentous networks in the extracellular matrices of various tissues. This protein contains two von Willebrand factor A domains; it is present in the cartilage extracellular matrix and has a role in the development and homeostasis of cartilage and bone. Mutations in this gene result in multiple epiphyseal dysplasia. [provided by RefSeq, Jul 2008]

Function:

Major component of the extracellular matrix of cartilage and may play a role in the formation of extracellular filamentous networks.

Subcellular Location:

Secreted.

Tissue Specificity:

Expressed only in cartilaginous tissues, such as vertebrae, ribs and shoulders.

DISEASE:

Defects in MATN3 are the cause of multiple epiphyseal dysplasia type 5 (EDM5) [MIM:607078]. EDM is a generalized skeletal dysplasia associated with significant morbidity. Joint pain, joint deformity, waddling gait, and short stature are the main clinical signs and symptoms. EDM is broadly categorized into the more severe Fairbank and the milder Ribbing types. EDM5 is relatively mild and clinically variable. It is primarily characterized by delayed and irregular ossification of the epiphyses and early-onset osteoarthritis.

Defects in MATN3 are the cause of spondyloepimetaphyseal dysplasia MATN3-related (SEMD-MATN3) [MIM:608728]. A bone disease characterized by disproportionate early-onset dwarfism, bowing of the lower limbs, lumbar lordosis and normal hands. Skeletal abnormalities include short, wide and stocky long bones with severe epiphyseal and metaphyseal changes, hypoplastic iliac bones and flat, ovoid vertebral bodies.

Genetic variations in MATN3 are associated with susceptibility to osteoarthritis type 2 (OS2) [MIM:140600]; also called osteoarthritis of distal interphalangeal joints (OADIP) or hand osteoarthritis (HOA). Osteoarthritis is a degenerative disease of the joints characterized by degradation of the hyaline articular cartilage and remodeling of the subchondral bone with sclerosis. Clinical symptoms include pain and joint stiffness often leading to significant disability and joint replacement. In the hand, osteoarthritis can develop in the distal interphalangeal and the first carpometacarpal (base of thumb) and proximal interphalangeal joints. Patients with osteoarthritis may have one, a few, or all of these sites affected.

Similarity:

Contains 4 EGF-like domains.

Contains 1 VWFA domain.

SWISS:

O15232

Gene ID:

4148

Important Note:

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