

Matrilin 3 Rabbit pAb

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Catalog # AP57217

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	O15232
Predicted	Rat, Pig, Cat, Dog, Human, Mouse, Rabbit, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	52817
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Matrilin 3
Epitope Specificity	51-150/486
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Secreted.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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]

Additional Information

Gene ID	4148
Other Names	Matrilin-3, MATN3
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:500 0-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	MATN3
Function	Major component of the extracellular matrix of cartilage and may play a role in the formation of extracellular filamentous networks.
Cellular Location	Secreted {ECO:0000250 UniProtKB:O35701}.
Tissue Location	Expressed only in cartilaginous tissues, such as vertebrae, ribs and shoulders

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