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LEPRE1 Rabbit pAb

Catalog # AP97626

Product Information

Application	IHC-P, IHC-F, IF, E
Primary Accession	Q32P28
Other Accession	Q32P28
Predicted	Rat, Pig, Human, Mouse, Rabbit, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	83394
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human LEPRE1
Epitope Specificity	351-450/736
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the leprecan family. Contains 1 Fe2OG dioxygenase domain. Contains 4 TPR repeats.
Post-translational modifications	O-glycosylated; chondroitin sulfate.
DISEASE	Defects in LEPRE1 are the cause of osteogenesis imperfecta type 8 (OI8) [MIM:610915]. A connective tissue disorder characterized by disproportionate short stature, severe osteoporosis, shortening of the long bones, white sclerae, a round face and a short barrel-shaped chest.
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 an enzyme that is a member of the collagen prolyl hydroxylase family. These enzymes are localized to the endoplasmic reticulum and their activity is required for proper collagen synthesis and assembly. Mutations in this gene are associated with osteogenesis imperfecta type VIII. Three alternatively spliced transcript variants encoding different isoforms have been described. Other variants may exist, but their biological validity has not been determined. [provided by RefSeq, Aug 2011]

Additional Information

Gene ID	64175
Other Names	GROS1; LEPRE1; OI8; 2410024C15Rik; P3H1_HUMAN; P3H1; Growth suppressor 1; Leucine- and proline-enriched proteoglycan 1 (Leprecan-1); 1.14.11.7; P3H1_MOUSE; P3H1_RAT; prolyl 3-hydroxylase 1; leucine proline-enriched proteoglycan (leprecan) 1; procollagen-proline 3-dioxygenase
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000

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	P3H1 (HGNC:19316)
Function	Has prolyl 3-hydroxylase activity and catalyzes the post- translational formation of 3-hydroxyproline in -Xaa-Pro-Gly- sequences in pro-collagen chains, a critical step for the formation of mature collagen trimers (PubMed:39245686). May be involved in the secretory pathway of cells. Has growth suppressive activity in fibroblasts.
Cellular Location	[Isoform 1]: Endoplasmic reticulum

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.