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PLOD2 Rabbit pAb

Catalog # AP98093

Product Information

Application	IHC-P, IHC-F, IF, E
Primary Accession	O00469
Other Accession	O00469
Predicted	Rat, Pig, Human, Mouse, Rabbit, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	84686
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human PLOD2
Epitope Specificity	501-600/737
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 1 Fe2OG dioxygenase domain.
DISEASE	Defects in PLOD2 are the cause of Bruck syndrome type 2 (BRKS2) [MIM:609220]. Bruck syndrome, also known as osteogenesis imperfecta with congenital joint contractures, is an autosomal recessive disease characterized by generalized osteopenia, joint contractures at birth, fragile bones and short stature. It can be distinguished from osteogenesis imperfecta by the absence of hearing loss and dentinogenesis imperfecta, and by the presence of clubfoot and congenital joint limitations. The molecular defect is an aberrant cross-linking of bone collagen, due to underhydroxylation of lysine residues within the telopeptides of type I collagen, whereas the lysine residues in the triple helix are normal.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The protein encoded by this gene is a membrane-bound homodimeric enzyme that is localized to the cisternae of the rough endoplasmic reticulum. The enzyme (cofactors iron and ascorbate) catalyzes the hydroxylation of lysyl residues in collagen-like peptides. The resultant hydroxylysyl groups are attachment sites for carbohydrates in collagen and thus are critical for the stability of intermolecular crosslinks. Some patients with Ehlers-Danlos syndrome type VIB have deficiencies in lysyl hydroxylase activity. Mutations in the coding region of this gene are associated with Bruck syndrome. Alternative splicing results in multiple transcript variants encoding different isoforms. [provided by RefSeq, Jul 2008]

Additional Information

Gene ID	5352
Other Names	BRKS2; LH2; TLH; D530025C14Rik; Plod-2; PLOD2_HUMAN; PLOD2; Lysyl hydroxylase 2 (LH2); 1.14.11.4; PLOD2_MOUSE; PLOD2_RAT;

	procollagen-lysine, 2-oxoglutarate 5-dioxygenase 2; procollagen-lysine, 2-oxoglutarate 5-dioxygenase (lysine hydroxylase) 2; procollagen-lysine, 2-oxoglutarate 5-dioxygenase 2; lysyl hydroxlase 2; telopeptidyl lysyl hydroxylase; procollagen lysyl hydroxylase 2
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	PLOD2 (HGNC:9082)
Function	Forms hydroxylysine residues in -Xaa-Lys-Gly- sequences in collagens. These hydroxylysines serve as sites of attachment for carbohydrate units and are essential for the stability of the intermolecular collagen cross-links.
Cellular Location	Rough endoplasmic reticulum membrane; Peripheral membrane protein; Lumenal side. Cytoplasm
Tissue Location	Highly expressed in pancreas and muscle. Isoform 1 and isoform 2 are expressed in the majority of the examined cell types Isoform 2 is specifically expressed in skin, lung, dura and aorta

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