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ENPP1 Recombinant Rabbit mAb

Catalog # AP98638

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P22413
Other Accession	P22413
Reactivity	Human
Host	Rabbit
Clonality	Recombinant
Calculated MW	104924
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human ENPP1
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	PBS, Glycerol, BSA.
SIMILARITY	Belongs to the peptidase S8 family. Furin subfamily.
SUBUNIT	Homodimer; disulfide-linked.
Post-translational modifications	Autophosphorylated as part of the catalytic cycle of phosphodiesterase/pyrophosphatase activity. N-glycosylated. It has been suggested that the active SMB domain may be permitted considerable disulfide bond heterogeneity or variability, thus two alternate disulfide patterns based on 3D structures are described with 1 disulfide bond conserved in both.
DISEASE	Defects in PCSK1 are the cause of proprotein convertase 1 deficiency (PC1 deficiency) [MIM:600955]. PC1 deficiency is characterized by obesity, hypogonadism, hypoadrenalism, reactive hypoglycemia as well as marked small-intestinal absorptive dysfunction It is due to impaired processing of prohormones.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	ENPP1 has a broad specificity and cleaves a variety of substrates, including phosphodiester bonds of nucleotides and nucleotide sugars and pyrophosphate bonds of nucleotides and nucleotide sugars. It can hydrolyze nucleoside 5' triphosphates such as ATP, GTP, CTP, TTP and UTP to their corresponding monophosphates with release of pyrophosphate. It can also hydrolyze diadenosine polyphosphates and 3',5'-cAMP to AMP. It may play a role in the regulation of pyrophosphate production, the regulation of the availability of nucleotide sugars in the endoplasmic reticulum and Golgi, and the regulation of purinergic signaling. The subtilisin-like Prohormone Convertase (PC) family is a group of cellular enzymes that cleave most prohormones and neuropeptide precursors. Numerous other cellular proteins, some viral proteins, and bacterial toxins that are transported by the constitutive secretory pathway are also targeted for maturation by PCs. PC family members share structural similarities, which include a heterogeneous ~10 kDa amino-terminal proregion, a highly conserved ~55 kDa subtilisin-like catalytic domain, and carboxyl-terminal domain that is heterogeneous in length and sequence. These enzymes become catalytically active following proregion cleavage within the appropriate cellular compartment. The

subcellular localization of PC family members varies. Immunolocalization studies show that PC1 is found in the perinuclear region as well as the trans-Golgi network, whereas PC2 can be found in the trans-Golgi network as well as diffusely distributed in the peripheral cytoplasm.

Additional Information

Gene ID 5167

Other Names ARHR2; COLED; M6S1; NPP1; NPPS; PC-1; PCA1; PDNP1; 4833416E15Rik; CD203c; E-NPP 1; E-NPP1; Ly-41; Pca; Pca-1; ttw; twy; Pc1; ENPP1_HUMAN; ENPP1; Alkaline phosphodiesterase I; Membrane component chromosome 6 surface marker 1; Nucleotide diphosphatase; Nucleotide pyrophosphatase (NPPase); Phosphodiesterase I/nucleotide pyrophosphatase 1; Plasma-cell membrane glycoprotein PC-1; ENPP1_MOUSE; Lymphocyte antigen 41 (Ly-41); ENPP1_RAT; ectonucleotide pyrophosphatase/phosphodiesterase 1

Dilution IHC-P=1:200-1000,IHC-F=1:200-1000,IF=1:200-1000

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 ENPP1 (HGNC:3356)

Function Nucleotide pyrophosphatase that generates diphosphate (PPi) and functions in bone mineralization and soft tissue calcification by regulating pyrophosphate levels (By similarity). PPi inhibits bone mineralization and soft tissue calcification by binding to nascent hydroxyapatite crystals, thereby preventing further growth of these crystals (PubMed:11004006). Preferentially hydrolyzes ATP, but can also hydrolyze other nucleoside 5' triphosphates such as GTP, CTP and UTP to their corresponding monophosphates with release of pyrophosphate, as well as diadenosine polyphosphates, and also 3',5'-cAMP to AMP (PubMed:25344812, PubMed:27467858, PubMed:28011303, PubMed:35147247, PubMed:8001561). May also be involved in the regulation of the availability of nucleotide sugars in the endoplasmic reticulum and Golgi, and the regulation of purinergic signaling (PubMed:27467858, PubMed:8001561). Inhibits ectopic joint calcification and maintains articular chondrocytes by repressing hedgehog signaling; it is however unclear whether hedgehog inhibition is direct or indirect (By similarity). Appears to modulate insulin sensitivity and function (PubMed:10615944). Also involved in melanogenesis (PubMed:28964717). Also able to hydrolyze 2',3'-cGAMP (cyclic GMP-AMP), a second messenger that activates TMEM173/STING and triggers type-I interferon production (PubMed:25344812). 2',3'-cGAMP degradation takes place in the lumen or extracellular space, and not in the cytosol where it is produced; the role of 2',3'-cGAMP hydrolysis is therefore unclear (PubMed:25344812). Not able to hydrolyze the 2',3'-cGAMP linkage isomer 3'-3'-cGAMP (PubMed:25344812).

Cellular Location [Ectonucleotide pyrophosphatase/phosphodiesterase family member 1]: Cell membrane; Single-pass type II membrane protein. Basolateral cell membrane; Single-pass type II membrane protein Note=Targeted to the basolateral membrane in polarized epithelial cells and in hepatocytes, and to matrix vesicles in osteoblasts (PubMed:11598187). In bile duct cells and

	cancer cells, located to the apical cytoplasmic side (PubMed:11598187)
Tissue Location	Expressed in plasma cells and also in a number of non-lymphoid tissues, including the distal convoluted tubule of the kidney, chondrocytes and epididymis (PubMed:9344668). Expressed in melanocytes but not in keratinocytes (PubMed:28964717)

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