

PCSK1/NEC1 Rabbit pAb

Catalog # AP97375

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P29120
Other Accession	P29120
Host	Rabbit
Clonality	Polyclonal
Calculated MW	84152
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human PCSK1/NEC1
Epitope Specificity	311-410/753
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 peptidase S8 family. Furin subfamily.
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	This gene encodes a member of the subtilisin-like proprotein convertase family, which includes proteases that process protein and peptide precursors trafficking through regulated or constitutive branches of the secretory pathway. The encoded protein undergoes an initial autocatalytic processing event in the ER to generate a heterodimer which exits the ER and sorts to subcellular compartments where a second autocatalytic even takes place and the catalytic activity is acquired. The protease is packaged into and activated in dense core secretory granules and expressed in the neuroendocrine system and brain. This gene encodes one of the seven basic amino acid-specific members which cleave their substrates at single or paired basic residues. It functions in the proteolytic activation of polypeptide hormones and neuropeptides precursors. Mutations in this gene have been associated with susceptibility to obesity and proprotein convertase 1/3 deficiency. Alternatively spliced transcript variants encoding multiple isoforms have been observed for this gene [provided by RefSeq, Jan 2014]

Additional Information

Gene ID	5122
Other Names	PC1; PC3; BMIQ12; NEC1; PC1/3; SPC3; Nec-1; Phpp-1; BDP; NEC1_BOVIN; PCSK1; NEC 1; Prohormone convertase 1; Proprotein convertase 1 (PC1); 3.4.21.93; NEC1_HUMAN; NEC1_MOUSE; Furin homolog;

Propeptide-processing protease; Att-1; NEC1_RAT; proprotein convertase subtilisin/kexin type 1; neuroendocrine convertase 1; prohormone convertase 3; proprotein convertase 1/3

Dilution

WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500

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

PCSK1

Synonyms

NEC1

Function

Involved in the processing of hormone and other protein precursors at sites comprised of pairs of basic amino acid residues. Substrates include POMC, renin, oxytocin, vasopressin, enkephalin, dynorphin, somatostatin, insulin and AGRP.

Cellular Location

Cytoplasmic vesicle, secretory vesicle. Note=Localized in the secretion granules

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