

PR3 Recombinant Rabbit mAb

Catalog # AP100070

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P24158
Other Accession	P24158
Host	Rabbit
Clonality	Recombinant
Calculated MW	27807
Physical State	Liquid
Immunogen	Recombinant human PR3 Protein
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 S1 family. Elastase subfamily. Contains 1 peptidase S1 domain.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The major features of Wegener granulomatosis are necrotizing granulomatous lesions, which most often affect the upper and lower airways and are associated with vasculitis, necrotizing glomerulonephritis and pulmonary capillaritis. The antigen responsible for this disease is Proteinase 3 (PR3, P29 or myeloblastin), which is one of the antibiotic proteins of neutrophilic granules belonging to the serine protease family. It is closely related to two others: Neutrophil Elastase and azurocidin. All three genes are expressed coordinately and their protein products are packaged together into azurophil granules during neutrophil differentiation. PR3 is a neutrophil protein which is able to cleave elastin and is involved in proliferation of human leukemia cells. PR3 is expressed specifically in immature myeloid cells and is a G-CSF-responsive protein critical to factor-independent growth. The genes for all three of the related serine protease family members are located in a cluster on the tip of the short arm of human chromosome 19.

Additional Information

Gene ID	5657
Other Names	ACPA; AGP7; C-ANCA; CANCA; MBN; MBT; NP-4; NP4; P29; PR-3; PR3; PRTN3_HUMAN; PRTN3; C-ANCA antigen; Leukocyte proteinase 3 (PR-3 PR3); Neutrophil proteinase 4 (NP-4); Wegener autoantigen; 3.4.21.76; proteinase 3; myeloblastin; serine proteinase, neutrophil; Wegener granulomatosis autoantigen
Dilution	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	PRTN3
Synonyms	MBN
Function	Serine protease that degrades elastin, fibronectin, laminin, vitronectin, and collagen types I, III, and IV (in vitro) (PubMed: 2033050 , PubMed: 28240246 , PubMed: 3198760). By cleaving and activating receptor F2RL1/PAR-2, enhances endothelial cell barrier function and thus vascular integrity during neutrophil transendothelial migration (PubMed: 23202369). Plays a role in neutrophil transendothelial migration, probably when associated with CD177 (PubMed: 22266279). Triggers inflammatory processes in neutrophils by interacting with ADGRG3 upstream of F2RL1/PAR2 activation (PubMed: 36302784).
Cellular Location	Cytoplasmic granule. Secreted. Cell membrane; Peripheral membrane protein; Extracellular side. Membrane raft; Peripheral membrane protein; Extracellular side. Note=Localizes predominantly to azurophil granules (primary secretory granules) in neutrophils (PubMed:18462208, PubMed:2033050, PubMed:3198760, PubMed:7897245) Secreted upon neutrophil stimulation by TNF, lipopolysaccharide (LPS), fMLP and CXCL8/IL8 or during neutrophil transmigration (PubMed:22266279, PubMed:28240246). Following secretion tethered to the cell membrane by CD177 (PubMed:18462208, PubMed:22266279)
Tissue Location	Expressed in polymorphonuclear leukocytes (at protein level) (PubMed:2033050, PubMed:3198760, PubMed:7897245) Expressed in neutrophils (at protein level) (PubMed:17244676, PubMed:18462208, PubMed:21193407, PubMed:22266279, PubMed:28240246) Expressed in differentiating neutrophils (PubMed:18462208)

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