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ELNE Recombinant Rabbit mAb

Catalog # AP100294

Product Information

Application	WB, IHC-P, IHC-F, IF, IP, E
Primary Accession	P08246
Other Accession	P08246
Reactivity	Human, Bovine
Host	Rabbit
Clonality	Recombinant
Calculated MW	28518
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.
SUBUNIT	Interacts with NOTCH2NL.
DISEASE	Defects in ELANE are a cause of cyclic haematopoiesis (CH) [MIM:162800]; also known as cyclic neutropenia. CH is an autosomal dominant disease in which blood-cell production from the bone marrow oscillates with 21-day periodicity. Circulating neutrophils vary between almost normal numbers and zero. During intervals of neutropenia, affected individuals are at risk for opportunistic infection. Monocytes, platelets, lymphocytes and reticulocytes also cycle with the same frequency. Defects in ELANE are the cause of neutropenia severe congenital autosomal dominant type 1 (SCN1) [MIM:202700]. SCN1 is a disorder of hematopoiesis characterized by a maturation arrest of granulopoiesis at the level of promyelocytes with peripheral blood absolute neutrophil counts below 0.5 x 10(9)/l and early onset of severe bacterial infections.
Background Descriptions	Elastases form a subfamily of serine proteases that hydrolyze many proteins in addition to elastin. Humans have six elastase genes which encode structurally similar proteins. The encoded preproprotein is proteolytically processed to generate the active protease. Following activation, this protease hydrolyzes proteins within specialized neutrophil lysosomes, called azurophil granules, as well as proteins of the extracellular matrix. The enzyme may play a role in degenerative and inflammatory diseases through proteolysis of collagen-IV and elastin. This protein also degrades the outer membrane protein A (OmpA) of E. coli as well as the virulence factors of such bacteria as Shigella, Salmonella and Yersinia. Mutations in this gene are associated with cyclic neutropenia and severe congenital neutropenia (SCN). This gene is present in a gene cluster on chromosome 19. [provided by RefSeq, Jan 2016

Additional Information

Gene ID	1991
Other Names	ELA2; GE; HLE; HNE; NE; PMN-E; SCN1; F430011M15Rik; ELNE_HUMAN; ELANE;

	Bone marrow serine protease; Elastase-2; Human leukocyte elastase (HLE); Medullasin; PMN elastase; 3.4.21.37; ELNE_MOUSE; Leukocyte elastase; elastase, neutrophil expressed; elastase 2, neutrophil; neutrophil elastase; polymorphonuclear leukocyte elastase; Neutrophil elastase (Bone marrow serine protease) (Elastase-2) (Human leukocyte elastase) (HLE) (Medullasin) (PMN elastase)
Dilution	IHC-P=1:50-200,IHC-F=1:50-200,WB=1:500-2000,IF=1:50-200,ELISA=1:5000-10000,IP=1:50-200
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	ELANE
Synonyms	ELA2
Function	Serine protease that modifies the functions of natural killer cells, monocytes and granulocytes (PubMed:15140022). Shows microbicidal activity in neutrophils by mediating cleavage and degradation of bacterial proteins (PubMed:10947984, PubMed:25161283). Capable of killing E.coli but not S.aureus in vitro; digests outer membrane protein A (ompA) in E.coli and K.pneumoniae (PubMed:10947984). Involved for the formation of neutrophil extracellular traps (NETs): translocates into the nucleus downstream of MPO and mediates degadation of histone H4, thereby participating to chromatin decondensation (PubMed:20974816, PubMed:25066128, PubMed:28574339). NETs are mainly composed of DNA fibers and are released by neutrophils to trap pathogens during inflammation (PubMed:20974816, PubMed:25066128, PubMed:28574339). Inhibits C5a-dependent neutrophil enzyme release and chemotaxis (PubMed:15140022). Promotes cleavage of GSDMB, thereby inhibiting pyroptosis (PubMed:36899106). Promotes blood coagulation (PubMed:20676107). Through the activation of the platelet fibrinogen receptor integrin alpha-IIb/beta-3, potentiates platelet aggregation induced by a threshold concentration of cathepsin G (CTSG) (PubMed:25211214, PubMed:9111081). Cleaves and thus inactivates tissue factor pathway inhibitor (TFPI) (PubMed:20676107, PubMed:25211214).
Cellular Location	Cytoplasmic vesicle, phagosome. Lysosome Cytolytic granule. Nucleus. Note=Localizes to azurophil granules, a type of primary lysosome found in neutrophils and monocytes that contains antimicrobial proteins (PubMed:20974816, PubMed:25066128) Translocates from azurophil granules to the nucleus downstream of MPO during formation of neutrophil extracellular traps (NETs), and is present in released NETs (PubMed:20974816, PubMed:25066128, PubMed:32434865). Localized in phagolysosomes following ingestion of E.coli by neutrophils (PubMed:10947984).
Tissue Location	Bone marrow cells. Neutrophil (PubMed:10947984).

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