中性粒细胞弹性蛋白酶 ELANE 抗体

产品货号： mIR23548

英文名称： Neutrophil Elastase

中文名称： 中性粒细胞弹性蛋白酶 ELANE 抗体

别　　名： Bone Marrow Serine Protease; ELA 2; ELA2; ELANE; Elastase 2; Elastase-2; Elastase 2 neutrophil; Elastase neutrophil expressed; Elastase-2; ELNE_HUMAN; GE antibody Granulocyte derived elastase; HLE; HNE; Human leukocyte elastase; Leukocyte elastase; Leukocyte Elastase Precursor; Medullasin; NE; Neutrophil elastase; Neutrophill elastase; PMN E; PMN Elastase; Polymorphonuclear elastase; SCN1; Serine protease; SERP1.

研究领域： 细胞生物　信号转导　细胞骨架　细胞外基质

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分 子 量： 26kDa

细胞定位： 细胞浆 细胞外基质

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human Neutrophil Elastase :51-150/267

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： Store at -20 ° C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20° C. 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.

PubMed : PubMed

产品介绍 : Elastases form a subfamily of serine proteases that hydrolyze many proteins in addition to elastin. Humans have six elastase genes which encode the structurally similar proteins. The product of this gene hydrolyzes proteins within specialized neutrophil lysosomes, called azurophil granules, as well as proteins of the extracellular matrix following the protein's release from activated neutrophils. The enzyme may play a role in degenerative and inflammatory diseases by its proteolysis of collagen-IV and elastin of the extracellular matrix. This protein 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 clustered with other serine protease gene family members, azurocidin 1 and proteinase 3 genes, at chromosome 19pter. All 3 genes are expressed coordinately and their protein products are packaged together into azurophil granules during neutrophil differentiation. [provided by RefSeq, May 2009].

Function:

Elastases form a subfamily of serine proteases that hydrolyze many proteins in addition to elastin. Humans have six elastase genes which encode the structurally similar proteins. The product of this gene hydrolyzes proteins within specialized neutrophil lysosomes, called azurophil granules, as well as proteins of the extracellular matrix following the protein's release from activated neutrophils. The enzyme may play a role in degenerative and inflammatory diseases by its proteolysis of collagen-IV and elastin of the extracellular matrix. This protein 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 clustered with other serine protease gene family members, azurocidin 1 and proteinase 3 genes, at chromosome 19pter. All 3 genes are expressed coordinately and their protein products are packaged together into azurophil granules during neutrophil differentiation. [provided by RefSeq, May 2009].

Subunit:

Interacts with NOTCH2NL.

Tissue Specificity:

Bone marrow cells.

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.

Similarity:

Belongs to the peptidase S1 family. Elastase subfamily. Contains 1 peptidase S1 domain.

SWISS:

P08246

Gene ID:

1991

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

产品图片