



Rabbit Anti-alpha 1 Antitrypsin antibody

SL0096R

Product Name:	alpha 1 Antitrypsin
Chinese Name:	α -1抗胰蛋白酶抗体
Alias:	A1-Antitrypsin; Tryptase; Alpha-1-Antitrypsin; alpha 1-antitrypsin; A1A; A1AT; AAT; Alpha 1 antiproteinase; Alpha 1 protease inhibitor; alpha-1-AT; alpha1 PI; alpha 1 proteinase inhibitor; Clade A (alpha 1 antiproteinase antitrypsin) member 1; MGC23330; MGC9222; PI; PI; PRO2275; Protease inhibitor 1 (anti elastase); Serpin A1; Serpin peptidase inhibitor clade A (alpha 1 antiproteinase antitrypsin) member 1; SerpinA1; A1AT_HUMAN; Alpha-1-antitrypsin; Alpha-1 protease inhibitor; Alpha-1-antiproteinase; Serpin A1.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	44kDa
Cellular localization:	Extracellular matrixSecretory protein
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human alpha 1 Antitrypsin:101-200/418
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	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
Product Detail:	Alpha-1-antitrypsin is a protease inhibitor, deficiency of which is associated with

emphysema and liver disease. The protein is encoded by a gene (PI) located on the distal long arm of chromosome 14.

Function:

Inhibitor of serine proteases. Its primary target is elastase, but it also has a moderate affinity for plasmin and thrombin. Irreversibly inhibits trypsin, chymotrypsin and plasminogen activator. The aberrant form inhibits insulin-induced NO synthesis in platelets, decreases coagulation time and has proteolytic activity against insulin and plasmin.

Short peptide from AAT (SPAAT) is a reversible chymotrypsin inhibitor. It also inhibits elastase, but not trypsin. Its major physiological function is the protection of the lower respiratory tract against proteolytic destruction by human leukocyte elastase (HLE).

Subcellular Location:

Secreted.

Short peptide from AAT: Secreted, extracellular space, extracellular matrix.

Tissue Specificity:

Plasma.

Post-translational modifications:

N-glycosylated. Differential glycosylation produces a number of isoforms. N-linked glycan at Asn-107 is alternatively di-antennary, tri-antennary or tetra-antennary. The glycan at Asn-70 is di-antennary with trace amounts of tri-antennary. Glycan at Asn-271 is exclusively di-antennary. Structure of glycans at Asn-70 and Asn-271 is Hex5HexNAc4. The structure of the antennae is Neu5Ac(alpha1-6)Gal(beta1-4)GlcNAc attached to the core structure Man(alpha1-6)[Man(alpha1-3)]Man(beta1-4)GlcNAc(beta1-4)GlcNAc. Some antennae are fucosylated, which forms a Lewis-X determinant.

Proteolytic processing may yield the truncated form that ranges from Asp-30 to Lys-418.

DISEASE:

Defects in SERPINA1 are the cause of alpha-1-antitrypsin deficiency (A1ATD) [MIM:613490]. A disorder whose most common manifestation is emphysema, which becomes evident by the third to fourth decade. A less common manifestation of the deficiency is liver disease, which occurs in children and adults, and may result in cirrhosis and liver failure. Environmental factors, particularly cigarette smoking, greatly increase the risk of emphysema at an earlier age.

Similarity:

Belongs to the serpin family.

SWISS:

P01009

Gene ID:

5265

Database links:

[Entrez Gene: 5265](#)Human

[Entrez Gene: 24648](#)Rat

[Omim: 107400](#)Human

[SwissProt: P01009](#)Human

[SwissProt: P07758](#)Mouse

[SwissProt: P17475](#)Rat

[Unigene: 525557](#)Human

Important Note:

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

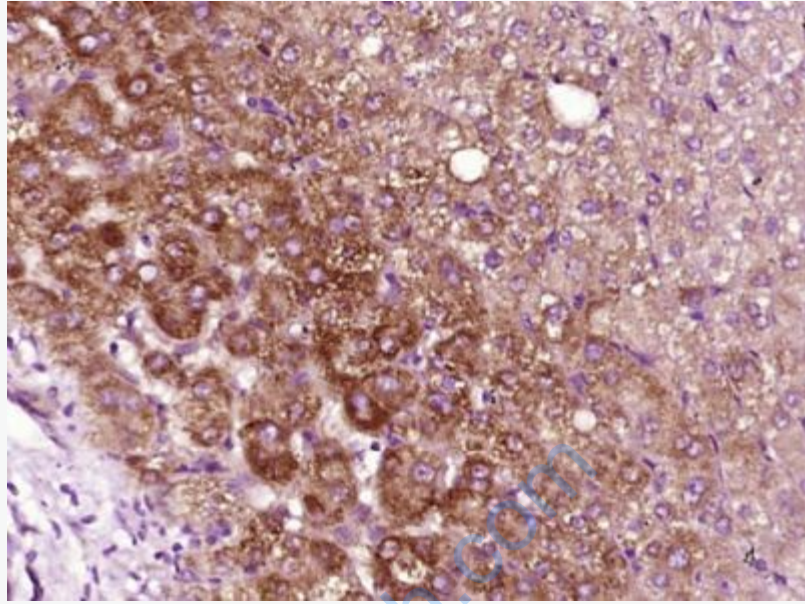
α 1-

抗胰蛋白酶(A1AT)缺乏是最常见的遗传代谢病,能引起肺和肝的损伤。 α 1抗胰蛋白酶为呼吸系统的非特异性可溶因子,与呼吸道抵抗力关系密切,它可抑制多种酶的活性,包括细菌的酶,以及中性白细胞溶酶体分泌的蛋白酶、弹性蛋白酶、胶原酶、纤维蛋白溶酶和凝血酶。 α 1抗胰蛋白酶的缺乏与慢性阻塞性肺病的形成关系密切,因为它的缺乏,不能及时控制感染和炎症产生的多种蛋白酶,而造成肺组织破坏。

α 1-抗胰蛋白酶由肝细胞产生,是一种分子量为45-

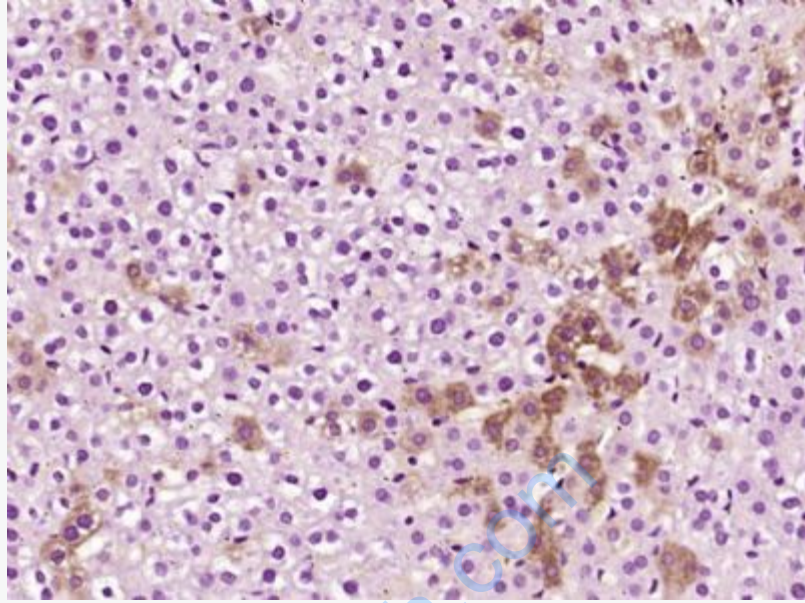
56kDa的glycoprotein,它能抑制蛋白酶、弹性蛋白酶、胶原酶等多种水解酶的活性。

AAT也是一种glycoprotein,主要用于遗传性AAT缺乏症和良性恶性肝Tumour、内胚窦瘤、组织细胞性淋巴瘤以及胰腺癌、胃癌、结肠癌等各种Tumour的研究。此抗体与大、小鼠、狗和猪有React Species。



Picture:

Paraformaldehyde-fixed, paraffin embedded (Human liver carcinoma); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (alpha 1 Antitrypsin) Polyclonal Antibody, Unconjugated (SL0096R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.



Paraformaldehyde-fixed, paraffin embedded (Rat liver); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (alpha 1 Antitrypsin) Polyclonal Antibody, Unconjugated (SL0096R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.