

AAT (H-203): sc-30121

BACKGROUND

Cumulative damage to lung tissue by neutrophil elastase is responsible for the development of pulmonary emphysema, an irreversible lung disease characterized by loss of lung elasticity. α 1-antitrypsin (AAT), a 394 amino-acid hepatic acute phase protein, predominantly inhibits neutrophil elastase. AAT is highly expressed in liver and in cultured hepatoma cells and, to a lesser extent, in macrophages. AAT is a highly polymorphic glycosylated serum protein with characteristic isoelectric-focusing patterns for most variants. AAT maps to a region of human chromosome 14q32.1 that includes a related serine protease inhibitor (serpin) gene which encodes corticosteroid-binding globulin. Oxidation of the Methionine 358 residue located at the active center of AAT results in a dramatic decrease in inhibitory activity towards elastase which effectively inactivates the protective function. AAT also has a moderate affinity for plasmin and Thrombin. AAT deficiency associates with a 20-30 fold increased risk of precocious pulmonary emphysema.

REFERENCES

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2. Seyama, K., Nukiwa, T., Takabe, K., Takahashi, H., Miyake, K. and Kira, S. 1991. Siyama (Serine 53 (TCC) to phenylalanine 53 (TTC)). A new α 1-antitrypsin-deficient variant with mutation on a predicted conserved residue of the serpin backbone. *J. Biol. Chem.* 266: 12627-12632.
3. Rosenberg, S., Barr, P.J., Najarian, R.C. and Hallelwell, R.A. 1994. Synthesis in yeast of a functional oxidation-resistant mutant of human α -antitrypsin. *Nature* 312: 77-80.
4. Graziadei, I., Vogel, W. and Bomford, A. 2000. A novel-binding site for the native hepatic acute-phase protein α -antitrypsin expressed on the human hepatoma cell line Hep G2 and intestinal cell line Caco 2. *Liver* 20: 240-246.
5. Rollini, P. and Fournier, R.E. 2000. Differential regulation of gene activity and chromatin structure within the human serpin gene cluster at 14q32.1 in macrophage microcell hybrids. *Nucleic Acids Res.* 28: 1767-1777.

CHROMOSOMAL LOCATION

Genetic locus: SERPINA1 (human) mapping to 14q32.13; Serpina1 (mouse) mapping to 12 E.

SOURCE

AAT (H-203) is a rabbit polyclonal antibody raised against amino acids 216-418 mapping at the C-terminus of AAT of human origin.

PRODUCT

Each vial contains 200 μ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

APPLICATIONS

AAT (H-203) is recommended for detection of AAT of human and, to a lesser extent, mouse and rat origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for AAT siRNA (h): sc-40945 and AAT siRNA (m): sc-40946.

Molecular Weight of luminal AAT: 51 kDa.

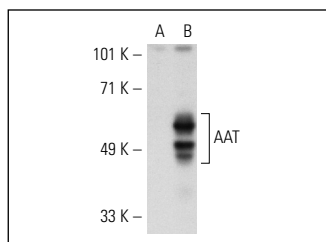
Molecular Weight of mature AAT: 55 kDa.

Positive Controls: human liver tissue extract or AAT (h): 293 Lysate: sc-112989.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



AAT (H-203): sc-30121. Western blot analysis of AAT expression in non-transfected: sc-110760 (A) and human AAT transfected: sc-112989 (B) 293 whole cell lysates.

RESEARCH USE

For research use only, not for use in diagnostic procedures.



Try **AAT (H-7): sc-166018** or **AAT (9): sc-130299**, our highly recommended monoclonal alternatives to AAT (H-203).