



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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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.
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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.
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CHROMOSOMAL LOCATION

Genetic locus: SERPINA1 (human) mapping to 14q32.13

SOURCE

AAT (703) is a mouse monoclonal antibody raised against purified AAT from serum of human origin.

STORAGE

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

PRODUCT

Each vial contains 100 μ g IgG₁ in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

AAT (703) is recommended for detection of α 1-antitrypsin (AAT) of human 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)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000); non cross-reactive with α 1-antichymotrypsin.

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

Molecular Weight of luminal AAT: 51 kDa.

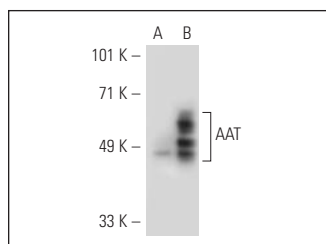
Molecular Weight of mature AAT: 55 kDa.

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

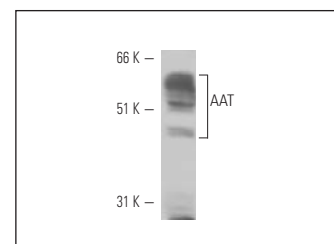
RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-mouse IgG-HRP: sc-2005 (dilution range: 1:2000-1:32,000) or Cruz Marker™ compatible goat anti-mouse IgG-HRP: sc-2031 (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).

DATA



AAT (703): sc-59435. Western blot analysis of AAT expression in non-transfected: sc-110760 (A) and human AAT transfected: sc-112989 (B) 293 whole cell lysates.



AAT (703): sc-59435. Western blot analysis of AAT expression in human liver tissue extract.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.