

## AAT (B9): sc-59438



The Power to Question

## 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.  $\alpha$ 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. The gene encoding AAT maps to a region of human chromosome 14q32.13 that includes a related serine protease inhibitor (serpin) gene which encodes corticosteroid-binding globulin. Oxidation of the methionine 358 residue in the active center of AAT results in a dramatic decrease in inhibitory activity towards elastase. AAT also has a moderate affinity for plasmin and Thrombin. AAT deficiency is associated with a 20-30 fold increased risk of precocious pulmonary emphysema.

## REFERENCES

- Okayama, H., et al. 1991. Characterization of the molecular basis of the  $\alpha$ 1-antitrypsin F allele. *Am. J. Hum. Genet.* 48: 1154-1158.
- Seyama, K., et al. 1991. Siiyama (Serine 53 (TCC) to phenylalanine 53 (TTC)). A new  $\alpha$ 1-antitrypsin-deficient variant with mutation on a predicted conserved residue of the serpin backbone. *J. Biol. Chem.* 266: 12627-12632.

## CHROMOSOMAL LOCATION

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

## SOURCE

AAT (B9) is a mouse monoclonal antibody raised against full length AAT of human origin.

## PRODUCT

Each vial contains 100  $\mu$ g IgG<sub>1</sub> in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

## APPLICATIONS

AAT (B9) is recommended for detection of AAT of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2  $\mu$ g per 100-500  $\mu$ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (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, AAT shRNA Plasmid (h): sc-40945-SH and AAT shRNA (h) Lentiviral Particles: sc-40945-V.

Molecular Weight of luminal AAT: 51 kDa.

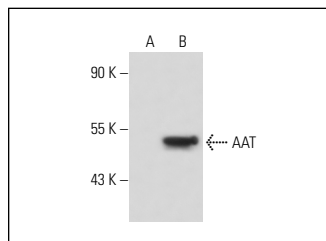
Molecular Weight of mature AAT: 55 kDa.

Positive Controls: human liver extract: sc-363766, Hep G2 cell lysate: sc-2227 or AAT (h): 293 Lysate: sc-112989.

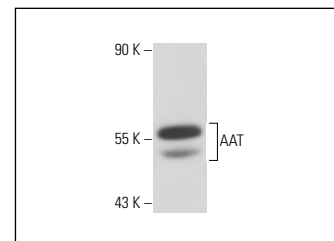
## STORAGE

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

## DATA



AAT (B9): sc-59438. Western blot analysis of AAT expression in non-transfected: sc-110760 (A) and human AAT transfected: sc-112989 (B) 293 whole cell lysates.



AAT (B9): sc-59438. Western blot analysis of AAT expression in human liver tissue extract.

## SELECT PRODUCT CITATIONS

- Janciauskiene, S., et al. 2008.  $\alpha$ 1-antitrypsin inhibits the activity of the matriptase catalytic domain *in vitro*. *Am. J. Respir. Cell Mol. Biol.* 39: 631-637.
- Subramaniam, D., et al. 2010. Cholesterol rich lipid raft microdomains are gateway for acute phase protein, SERPINA1. *Int. J. Biochem. Cell Biol.* 42: 1562-1570.
- Burkard, A., et al. 2012. Generation of proliferating human hepatocytes using Upcyte® technology: characterisation and applications in induction and cytotoxicity assays. *Xenobiotica* 42: 939-956.
- Ferrarotti, I., et al. 2015. How can we improve the detection of  $\alpha$ 1-antitrypsin deficiency? *PLoS ONE* 10: e0135316.
- Izquierdo, I., et al. 2016. Proteomic identification of putative biomarkers for early detection of sudden cardiac death in a family with a LMNA gene mutation causing dilated cardiomyopathy. *J. Proteomics* 148: 75-84.
- Pastore, N., et al. 2017. Activation of the c-Jun N-terminal kinase pathway aggravates proteotoxicity of hepatic mutant Z  $\alpha$ 1-antitrypsin. *Hepatology* 65: 1865-1874.
- Matamala, N., et al. 2018. Characterization of novel missense variants of SERPINA1 gene causing  $\alpha$ -1 antitrypsin deficiency. *Am. J. Respir. Cell Mol. Biol.* 58: 706-716.
- Kim, E.K., et al. 2019. Proteomic analysis of primary colon cancer and synchronous solitary liver metastasis. *Cancer Genomics Proteomics* 16: 583-592.
- Gómez-Mariano, G., et al. 2020. Liver organoids reproduce  $\alpha$ 1 antitrypsin deficiency-related liver disease. *Hepatol. Int.* 14: 127-137.

## RESEARCH USE

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