SANTA CRUZ BIOTECHNOLOGY, INC.

AAT (B9): sc-59438

SANTA CRUZ BIOTECHNOLOGY

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. 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. 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

1. Okayama, H., et al. 1991. Characterization of the molecular basis of the α1-antitrypsin F allele. Am. J. Hum. Genet. 48: 1154-1158.
2. Seyama, K., et al. 1991. Siiyama (Serine 53 (TCC) to phenylalanine 53 (TTC)). A new α1-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 µg IgG_1 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 µg per 100-500 µg 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.

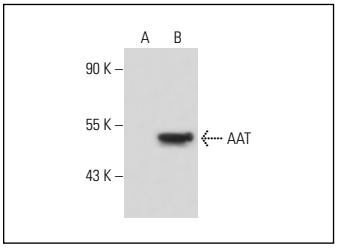
Molecular Weight of mature AAT: 55 kDa.

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

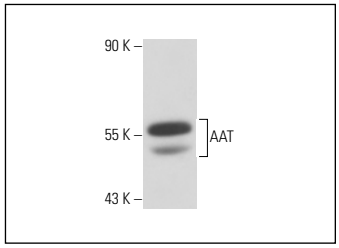
STORAGE

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

1. Janciauskiene, S., et al. 2008. α1-antitrypsin inhibits the activity of the matriptase catalytic domain *in vitro*. Am. J. Respir. Cell Mol. Biol. 39: 631-637.
2. Subramaniyam, D., et al. 2010. Cholesterol rich lipid raft microdomains are gateway for acute phase protein, SERPINA1. Int. J. Biochem. Cell Biol. 42: 1562-1570.
3. 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.
4. Ferrarotti, I., et al. 2015. How can we improve the detection of α1-antitrypsin deficiency? PLoS ONE 10: e0135316.
5. 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.
6. Pastore, N., et al. 2017. Activation of the c-Jun N-terminal kinase pathway aggravates proteotoxicity of hepatic mutant Z α1-antitrypsin. Hepatology 65: 1865-1874.
7. Matamala, N., et al. 2018. Characterization of novel missense variants of SERPINA1 gene causing α-1 antitrypsin deficiency. Am. J. Respir. Cell Mol. Biol. 58: 706-716.
8. Kim, E.K., et al. 2019. Proteomic analysis of primary colon cancer and synchronous solitary liver metastasis. Cancer Genomics Proteomics 16: 583-592.
9. Gómez-Mariano, G., et al. 2020. Liver organoids reproduce α1 antitrypsin deficiency-related liver disease. Hepatol. Int. 14: 127-137.

RESEARCH USE

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