SANTA CRUZ BIOTECHNOLOGY, INC.

AGA (H-300): sc-98735

BACKGROUND

AGA (aspartylglucosaminidase) is a 346 amino acid precursor protein that belongs to the Ntn-hydrolase family and is cleaved to produce an α chain and a β chain. Localized to the lysosome, AGA functions as a heterotetramer composed of two α and two β chains that work together to cleave the GlcNAc-Asn bond that joins oligosaccharides to target glycoproteins. Defects in the gene encoding AGA are the cause of aspartylglucosaminuria (AGU), a lysosomal storage disease that is characterized by severe mental retardation and mild connective tissue abnormalities. The gene encoding AGA maps to human chromosome 4, which encodes nearly 6% of the human genome and has the largest gene deserts (regions of the genome with no protein encoding genes) of all of the human chromosomes.

REFERENCES

1. Mononen, I., Fisher, K.J., Kaartinen, V. and Aronson, N.N. 1993. Aspartylglycosaminuria: protein chemistry and molecular biology of the most common lysosomal storage disorder of glycoprotein degradation. FASEB J. 7: 1247-1256.
2. Tollersrud, O.K., Heiskanen, T. and Peltonen, L. 1994. Human leucocyte glycosylasparaginase is an α/β-heterodimer of 19 kDa α subunit and 17 and 18 kDa β subunit. Biochem. J. 300: 541-544.
3. Saarela, J., Laine, M., Oinonen, C., Schantz, C., Jalanko, A., Rouvinen, J. and Peltonen, L. 2001. Molecular pathogenesis of a disease: structural consequences of aspartylglucosaminuria mutations. Hum. Mol. Genet. 10: 983-995.
4. Saarela, J., Oinonen, C., Jalanko, A., Rouvinen, J. and Peltonen, L. 2004. Autoproteolytic activation of human aspartylglucosaminidase. Biochem. J. 378: 363-371.
5. Saarela, J., von Schantz, C., Peltonen, L. and Jalanko, A. 2004. A novel aspartylglucosaminuria mutation affects translocation of aspartylglucosaminidase. Hum. Mutat. 24: 350-351.
6. Jackson, M., Clayton, P., Grunewald, S., Keir, G., Mills, K., Mills, P., Winchester, B., Worthington, V. and Young, E. 2005. Elevation of plasma aspartylglucosaminidase is a useful marker for the congenital disorders of glycosylation type I (CDG I). J. Inherit. Metab. Dis. 28: 1197-1198.
7. Saito, S., Ohno, K., Sugawara, K., Suzuki, T., Togawa, T. and Sakuraba, H. 2008. Structural basis of aspartylglucosaminuria. Biochem. Biophys. Res. Commun. 377: 1168-1172.
8. Michelakakis, H., Moraitou, M., Mavridou, I. and Dimitriou, E. 2009. Plasma lysosomal enzyme activities in congenital disorders of glycosylation, galactosemia and fructosemia. Clin. Chim. Acta 401: 81-83.

CHROMOSOMAL LOCATION

Genetic locus: AGA (human) mapping to 4q34.3; Aga (mouse) mapping to 8 B1.3.

SOURCE

AGA (H-300) is a rabbit polyclonal antibody raised against amino acids 47-272 mapping at the C-terminus of AGA of human origin.

PRODUCT

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

APPLICATIONS

AGA (H-300) is recommended for detection of AGA of mouse, rat and 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

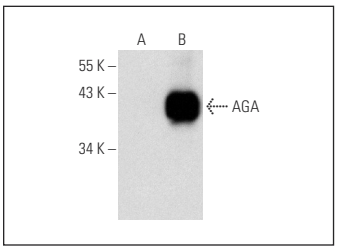
AGA (H-300) is also recommended for detection of AGA in additional species, including canine and porcine.

Suitable for use as control antibody for AGA siRNA (h): sc-89013, AGA siRNA (m): sc-105048, AGA shRNA Plasmid (h): sc-89013-SH, AGA shRNA Plasmid (m): sc-105048-SH, AGA shRNA (h) Lentiviral Particles: sc-89013-V and AGA shRNA (m) Lentiviral Particles: sc-105048-V.

Molecular Weight of AGA precursor: 39 kDa.

Positive Controls: AGA (h3): 293T Lysate: sc-112982.

DATA



AGA (H-300): sc-98735. Western blot analysis of AGA expression in non-transfected: sc-117752 (**A**) and human AGA transfected: sc-112982 (**B**) 293T whole cell lysates.

STORAGE

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

RESEARCH USE

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

MONOS Satisfaction Guaranteed

Try **AGA (H-8): sc-514075** or **AGA (G-10): sc-365848**, our highly recommended monoclonal alternatives to AGA (H-300).