SANTA CRUZ BIOTECHNOLOGY, INC.

AGA (G-10): sc-365848

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., et al. 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., et al. 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., et al. 2001. Molecular pathogenesis of a disease: structural consequences of aspartylglucosaminuria mutations. Hum. Mol. Genet. 10: 983-995.
4. Saarela, J., et al. 2004. Autoproteolytic activation of human aspartylglucosaminidase. Biochem. J. 378: 363-371.
5. Saarela, J., et al. 2004. A novel aspartylglucosaminuria mutation affects translocation of aspartylglucosaminidase. Hum. Mutat. 24: 350-351.
6. Jackson, M., et al. 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., et al. 2008. Structural basis of aspartylglucosaminuria. Biochem. Biophys. Res. Commun. 377: 1168-1172.
8. Michelakakis, H., et al. 2009. Plasma lysosomal enzyme activities in congenital disorders of glycosylation, galactosemia and fructosemia. Clin. Chim. Acta 401: 81-83.
9. Online Mendelian Inheritance in Man, OMIM™. 2009. Johns Hopkins University, Baltimore, MD. MIM Number: 208400. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/

CHROMOSOMAL LOCATION

Genetic locus: AGA (human) mapping to 4q34.3.

SOURCE

AGA (G-10) is a mouse monoclonal antibody raised against amino acids 47-272 mapping at the C-terminus of AGA of human origin.

RESEARCH USE

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

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

AGA (G-10) is recommended for detection of AGA of human origin by Western Blotting (starting dilution 1:100, 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).

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

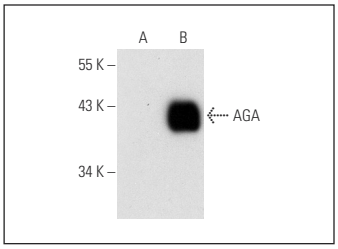
Molecular Weight of AGA precursor: 39 kDa.

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

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 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 m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



AGA (G-10): sc-365848. 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.

PROTOCOLS

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