SANTA CRUZ BIOTECHNOLOGY, INC.

AGA (H-8): sc-514075

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.

CHROMOSOMAL LOCATION

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

SOURCE

AGA (H-8) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 160-181 within an internal region of AGA of human origin.

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.

AGA (H-8) is available conjugated to agarose (sc-514075 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-514075 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-514075 PE), fluorescein (sc-514075 FITC), Alexa Fluor® 488 (sc-514075 AF488), Alexa Fluor® 546 (sc-514075 AF546), Alexa Fluor® 594 (sc-514075 AF594) or Alexa Fluor® 647 (sc-514075 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-514075 AF680) or Alexa Fluor® 790 (sc-514075 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

Blocking peptide available for competition studies, sc-514075 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

APPLICATIONS

AGA (H-8) is recommended for detection of AGA precursor and α chain of mouse, rat and 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 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.

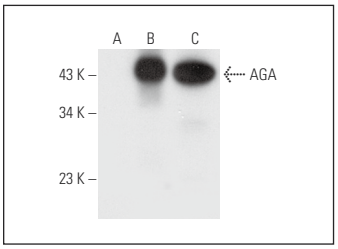
Molecular Weight of AGA α chain: 24 kDa.

Positive Controls: Jurkat whole cell lysate: sc-2204, AGA (h3): 293T Lysate: sc-112982 or human heart extract: sc-363763.

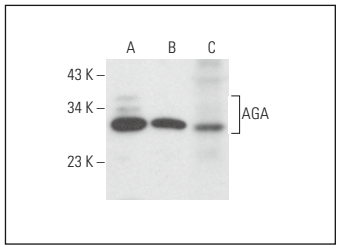
RECOMMENDED SUPPORT REAGENTS

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 (H-8): sc-514075. Western blot analysis of AGA expression in non-transfected: sc-117752 (**A**) and human AGA transfected: sc-112982 (**B**) 293T whole cell lysates and human heart tissue extract (**C**).



AGA (H-8): sc-514075. Western blot analysis of AGA expression in Jurkat (**A**) and ALL-SIL (**B**) whole cell lysates and rat lung tissue extract (**C**).

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.