SANTA CRUZ BIOTECHNOLOGY, INC.

α-gal A (C6): sc-517442

BACKGROUND

α-galactosidase A (α-gal A) functions as a lysosomal hydrolase. α-gal A forms an active homodimer that acts upon a glycolipid substrate, globotriaosylceramide (Gb3). Inherited mutations in the gene encoding α-gal A cause an X-linked recessive glycolipid storage disorder known as Fabry's disease. In Fabry patients, α-gal A deficiencies lead to an accumulation of Gb3 in the body. The numerous clinical manifestations of the disease include renal and cardiac impairment, severe pain in the extremities and cutaneous lesions known as angiokeratomas. Enzyme replacement therapy using recombinant α-gal A effectively treats the symptoms of Fabry disease.

REFERENCES

1. Kint, J.A. 1970. Fabry's disease: α-galactosidase deficiency. Science 167: 1268-1269.
2. Sweatman, A.K., et al. 1994. Physical mapping in the region of the Bruton's tyrosine kinase and α-galactosidase A gene loci in proximal Xq22. Hum. Genet. 94: 624-628.
3. Schiffmann, R., et al. 2000. Infusion of α-galactosidase A reduces tissue globotriaosylceramide storage in patients with Fabry disease. Proc. Natl. Acad. Sci. USA 97: 365-370.
4. Ioannou, Y.A., et al. 2001. Fabry disease: preclinical studies demonstrate the effectiveness of α-galactosidase A replacement in enzyme-deficient mice. Am. J. Hum. Genet. 68: 14-25.
5. Eng, C.M., et al. 2001. A phase 1/2 clinical trial of enzyme replacement in Fabry disease: pharmacokinetic, substrate clearance, and safety studies. Am. J. Hum. Genet. 68: 711-722.
6. Breunig, F., et al. 2003. Fabry disease: diagnosis and treatment. Kidney Int. Suppl. 84: 181-185.

CHROMOSOMAL LOCATION

Genetic locus: GLA (human) mapping to Xq22.1.

SOURCE

α-gal A (C6) is a mouse monoclonal antibody raised against a recombinant protein corresponding to amino acids 81-429 of α-gal A of human origin.

PRODUCT

Each vial contains 100 µg IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide, 0.1% gelatin and 1% glycerol.

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.

APPLICATIONS

α-gal A (C6) is recommended for detection of α-gal A 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 α-gal A siRNA (h): sc-105019, α-gal A shRNA Plasmid (h): sc-105019-SH and α-gal A shRNA (h) Lentiviral Particles: sc-105019-V.

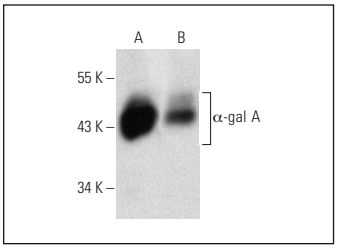
Molecular Weight of α-gal A: 50 kDa.

Positive Controls: NCI-H460 whole cell lysate: sc-364235.

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. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



α-gal A (C6): sc-517442. Western blot analysis of α-gal A expression in 293T (**A**) and NCI-H460 (**B**) whole cell lysates.

RESEARCH USE

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