SANTA CRUZ BIOTECHNOLOGY, INC.

α-gal A (N-16)-R: sc-26079-R

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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; Gla (mouse) mapping to X E-F1.

SOURCE

α-gal A (N-16)-R is an affinity purified rabbit polyclonal antibody raised against a peptide mapping near the N-terminus of α-galactosidase A 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.

Blocking peptide available for competition studies, sc-26079 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

α-gal A (N-16)-R is recommended for detection of α-gal A of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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).

α-gal A (N-16) is also recommended for detection of α-gal A in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for α-gal A siRNA (h): sc-105019, α-gal A siRNA (m): sc-140596, α-gal A shRNA Plasmid (h): sc-105019-SH, α-gal A shRNA Plasmid (m): sc-140596-SH, α-gal A shRNA (h) Lentiviral Particles: sc-105019-V and α-gal A shRNA (m) Lentiviral Particles: sc-140596-V.

Molecular Weight of α-gal A: 50 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

RESEARCH USE

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

PROTOCOLS

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