SANTA CRUZ BIOTECHNOLOGY, INC.

α-gal A (H-104): sc-25823

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). The gene encoding α-gal A maps to chromosome Xq22.1. Inherited mutations in this gene 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 $G_{\beta 3}$ 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.

CHROMOSOMAL LOCATION

Genetic locus: GLA (human) mapping to Xq22.1; Gla (mouse) mapping to X E3.

SOURCE

α-gal A (H-104) is a rabbit polyclonal antibody raised against amino acids 326-429 mapping at the C-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.

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.

PROTOCOLS

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

APPLICATIONS

α-gal A (H-104) is recommended for detection of α-gal A of human and, to a lesser extent, mouse and rat 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 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.

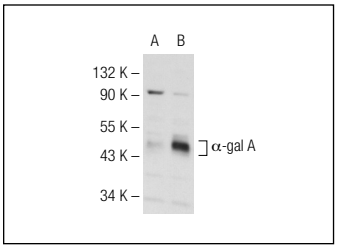
Molecular Weight of α-gal A: 50 kDa.

Positive Controls: α-gal A (h): 293T Lysate: sc-159292.

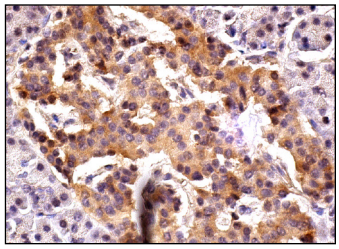
RECOMMENDED SECONDARY REAGENTS

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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) 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. 4) Immunohistochemistry: use ImmunoCruz™: sc-2051 or ABC: sc-2018 rabbit IgG Staining Systems.

DATA



α-gal A (H-104): sc-25823. Western blot analysis of α-gal A expression in non-transfected: sc-117752 (**A**) and human α-gal A transfected: sc-159292 (**B**) 293T whole cell lysates.



α-gal A (H-104): sc-25823. Immunoperoxidase staining of formalin fixed, paraffin-embedded human pancreas tissue showing cytoplasmic staining of Islets of Langerhans.

SELECT PRODUCT CITATIONS

1. Maalouf, K., et al. 2010. A modified lipid composition in Fabry disease leads to an intracellular block of the detergent-resistant membrane-associated dipeptidyl peptidase IV. J. Inherit. Metab. Dis. 33: 445-449.
2. Corchero, J.L., et al. 2011. Integrated approach to produce a recombinant, His-tagged human α-galactosidase A in mammalian cells. Biotechnol. Prog. 27: 1206-1217.