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Lieferung & Zahlungsart

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- Gefahrgutzuschlag
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SANTA CRUZ BIOTECHNOLOGY, INC.

α-gal A (h): 293T Lysate: sc-159292

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 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

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CHROMOSOMAL LOCATION

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

PRODUCT

α-gal A (h): 293T Lysate represents a lysate of human α-gal A transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

APPLICATIONS

α-gal A (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive α-gal A antibodies. Recommended use: 10-20 µl per lane.

Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

RESEARCH USE

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

PROTOCOLS

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