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Lieferung & Zahlungsart

siehe unsere Liefer- und Versandbedingungen

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- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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SANTA CRUZ BIOTECHNOLOGY, INC.

Neu1 (h): 293T Lysate: sc-171286

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

NEU1 encodes the lysosomal enzyme neuraminidase, Neu1, which cleaves terminal sialic acid residues from substrates such as glycoproteins and glycolipids. In the lysosome Neu1 belongs to a heterotrimeric complex containing β-galactosidase and cathepsin A (also referred to as "protective protein"). In humans, primary or secondary deficiency of this enzyme leads to two clinically similar neurodegenerative lysosomal storage disorders: sialidosis and galactosialidosis (GS). Sialidosis symptoms range from eye abnormalities and neurological disturbances to skeletal malformations, mental retardation and early death. Neu1 is expressed in the pancreas, muscle, kidney, placenta, heart, lung and liver. The human Neu1 gene maps to chromosome 6p21.33 and encodes a lysosomal protein localized on the inner side of the plasma membrane and in intracellular vesicles. Neu1 is also known as α-N-acetylneuraminidase and acetylneuraminyl hydrolase.

REFERENCES

1. Penzel, R., Uhl, J., Kopitz, J., Beck, M., Otto, H.F. and Cantz, M. 2001. Splice donor site mutation in the lysosomal neuraminidase gene causing exon skipping and complete loss of enzyme activity in a sialidosis patient. FEBS Lett. 501: 135-138.
2. Sergi, C., Penzel, R., Uhl, J., Zoubaa, S., Dietrich, H., Decker, N., Rieger, P., Kopitz, J., Otto, H.F., Kiessling, M. and Cantz, M. 2001. Prenatal diagnosis and fetal pathology in a Turkish family harboring a novel nonsense mutation in the lysosomal α-N-acetyl-neuraminidase (sialidase) gene. Hum. Genet. 109: 421-428.
3. de Geest, N., Bonten, E., Mann, L., de Sousa-Hitzler, J., Hahn, C. and d'Azzo, A. 2002. Systemic and neurologic abnormalities distinguish the lysosomal disorders sialidosis and galactosialidosis in mice. Hum. Mol. Genet. 11: 1455-1464.
4. Uhl, J., Penzel, R., Sergi, C., Kopitz, J., Otto, H.F. and Cantz, M. 2002. Identification of a CTL4/Neu1 fusion transcript in a sialidosis patient. FEBS Lett. 521: 19-23.
5. LocusLink Report (LocusID: 4758). http://www.ncbi.nlm.nih.gov/LocusLink/

CHROMOSOMAL LOCATION

Genetic locus: NEU1 (human) mapping to 6p21.33.

PRODUCT

Neu1 (h): 293T Lysate represents a lysate of human Neu1 transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

Neu1 (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive Neu1 antibodies. Recommended use: 10-20 µl per lane.

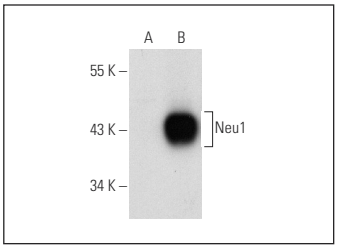
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

Neu1 (F-8): sc-166824 is recommended as a positive control antibody for Western Blot analysis of enhanced human Neu1 expression in Neu1 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

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.

DATA



Neu1 (F-8): sc-166824. Western blot analysis of Neu1 expression in non-transfected: sc-117752 (**A**) and human Neu1 transfected: sc-171286 (**B**) 293T whole cell lysates.

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.

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.