

# Neu1 (C-20): sc-25907

## 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  $\beta$ -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  $\alpha$ -N-acetylneuraminidase and Acetylneuraminyl hydrolase.

## REFERENCES

1. Sergi, C., et al. 2001. Prenatal diagnosis and fetal pathology in a Turkish family harboring a novel nonsense mutation in the lysosomal  $\alpha$ -N-acetylneuraminidase (sialidase) gene. *Hum. Genet.* 109: 421-428.
2. Penzel, R., et al. 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.
3. de Geest, N., et al. 2002. Systemic and neurologic abnormalities distinguish the lysosomal disorders sialidosis and galactosialidosis in mice. *Hum. Mol. Genet.* 11: 1455-1464.
4. Uhl, J., et al. 2002. Identification of a CTL4/Neu1 fusion transcript in a sialidosis patient. *FEBS Lett.* 521: 19-23.
5. SWISS-PROT/TrEMBL (17368612). World Wide Web URL: <http://www.expasy.ch/sprot/sprot-top.html>
6. LocusLink Report (LocusID: 4758). <http://www.ncbi.nlm.nih.gov/LocusLink/>

## CHROMOSOMAL LOCATION

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

## SOURCE

Neu1 (C-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of Neu1 of human origin.

## PRODUCT

Each vial contains 200  $\mu$ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-25907 P, (100  $\mu$ g 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

Neu1 (C-20) is recommended for detection of Neu1 of 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).

Neu1 (C-20) is also recommended for detection of Neu1 in additional species, including canine and porcine.

Suitable for use as control antibody for Neu1 siRNA (h): sc-106297, Neu1 shRNA Plasmid (h): sc-106297-SH and Neu1 shRNA (h) Lentiviral Particles: sc-106297-V.

Molecular Weight of Neu1: 45 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (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 donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (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](http://www.scbt.com) or our catalog for detailed protocols and support products.


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