SANTA CRUZ BIOTECHNOLOGY, INC.

alpha-L-iduronidase (L-12): sc-241189

BACKGROUND

Alpha-L-iduronidase, also known as IDUA (iduronidase, alpha-L-) or MPS1, is a 653 amino acid protein belonging to the glycosyl hydrolase 39 family. Encoded by a gene that maps to human chromosome 4p16.3, alpha-L-iduronidase localizes to lysosome and is ubiquitously expressed. Alpha-L-iduronidase hydrolyzes terminal alpha-L-iduronic acid residues of two glycosaminoglycans, dermatan sulfate and heparan sulfate, thereby resulting in lysosomal degradation of both glycosaminoglycans. Alpha-L-iduronidase defects are linked to mucopolysaccharidosis type 1H (MPS1H), also known as Hurler syndrome, a rare lysosomal storage disease characterized by progressive physical deterioration, urinary excretion of dermatan sulfate and heparan sulfate, hepatosplenomegaly, skeletal deformities, corneal clouding, severe mental retardation, obstructive airways disease, respiratory infection and cardiac complications.

REFERENCES

1. Scott, H.S., et al. 1991. Human alpha-L-iduronidase: cDNA isolation and expression. Proc. Natl. Acad. Sci. USA 88: 9695-9699.
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3. Scott, H.S., et al. 1992. alpha-L-iduronidase mutations (Q70X and P533R) associate with a severe Hurler phenotype. Hum. Mutat. 1: 333-339.
4. Bach, G., et al. 1993. Molecular analysis of Hurler syndrome in Druze and Muslim Arab patients in Israel: multiple allelic mutations of the IDUA gene in a small geographic area. Am. J. Hum. Genet. 53: 330-338.
5. Scott, H.S., et al. 1993. Identification of mutations in the alpha-L-iduronidase gene (IDUA) that cause Hurler and Scheie syndromes. Am. J. Hum. Genet. 53: 973-986.
6. Clarke, L.A. and Scott, H.S. 1993. Two novel mutations causing mucopolysaccharidosis type I detected by single strand conformational analysis of the alpha-L-iduronidase gene. Hum. Mol. Genet. 2: 1311-1312.
7. Scott, H.S., et al. 1993. Multiple polymorphisms within the alpha-L-iduronidase gene (IDUA): implications for a role in modification of MPS-I disease phenotype. Hum. Mol. Genet. 2: 1471-1473.
8. Tieu, P.T., et al. 1995. Four novel mutations underlying mild or intermediate forms of alpha-L-iduronidase deficiency (MPS IS and MPS IH/S). Hum. Mutat. 6: 55-59.
9. Brooks, D.A., et al. 2001. Glycosidase active site mutations in human alpha-L-iduronidase. Glycobiology 11: 741-750.

CHROMOSOMAL LOCATION

Genetic locus: IDUA (human) mapping to 4p16.3; Idua (mouse) mapping to 5 F.

SOURCE

alpha-L-iduronidase (L-12) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of alpha-L-iduronidase 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-241189 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

alpha-L-iduronidase (L-12) is recommended for detection of alpha-L-iduronidase 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).

alpha-L-iduronidase (L-12) is also recommended for detection of alpha-L-iduronidase in additional species, including canine.

Suitable for use as control antibody for alpha-L-iduronidase siRNA (h): sc-89031, alpha-L-iduronidase siRNA (m): sc-141030, alpha-L-iduronidase shRNA Plasmid (h): sc-89031-SH, alpha-L-iduronidase shRNA Plasmid (m): sc-141030-SH, alpha-L-iduronidase shRNA (h) Lentiviral Particles: sc-89031-V and alpha-L-iduronidase shRNA (m) Lentiviral Particles: sc-141030-V.

Molecular Weight of alpha-L-iduronidase: 74 kDa.

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.

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.