



MANBA siRNA (m): sc-149247

BACKGROUND

MANBA (mannosidase, β A, lysosomal), also known as mannanase or MANB1, is an 879 amino acid lysosomal exoglycosidase that cleaves the non-reducing ends of β -linked mannose residues of N-linked glycoprotein oligosaccharides. A member of the glycosyl hydrolase 2 family, MANBA is encoded by a gene that maps to human chromosome 4q24. Defects in the MANBA gene are the cause of an autosomal recessive lysosomal storage disease known as lysosomal β -mannosidosis (LYSBMAN). Patients with LSBMAN may exhibit various clinical features, but most cases are characterized by mental retardation, and sometimes hypotonia, epilepsy, speech impairment, hearing loss or peripheral neuropathy.

REFERENCES

- Cooper, A., Sardharwalla, I.B. and Roberts, M.M. 1986. Human β -mannosidase deficiency. N. Engl. J. Med. 315: 1231.
- Poenaru, L., Akli, S., Rocchiccioli, F., Eydoux, P. and Zamet, P. 1992. Human β -mannosidosis: a 3-year-old boy with speech impairment and emotional instability. Clin. Genet. 41: 331-334.
- Alkhaty, A.H., Kraemer, S.A., Leipprandt, J.R., Macek, M., Kleijer, W.J. and Friderici, K.H. 1998. Human β -mannosidase cDNA characterization and first identification of a mutation associated with human β -mannosidosis. Hum. Mol. Genet. 7: 75-83.
- Bedilu, R., Nummy, K.A., Cooper, A., Wevers, R., Smeitink, J., Kleijer, W.J. and Friderici, K.H. 2002. Variable clinical presentation of lysosomal β -mannosidosis in patients with null mutations. Mol. Genet. Metab. 77: 282-290.
- Uchino, Y., Fukushima, T., Yotsumoto, S., Hashiguchi, T., Taguchi, H., Suzuki, N., Konohana, I. and Kanzaki, T. 2003. Morphological and biochemical studies of human β -mannosidosis: identification of a novel β -mannosidase gene mutation. Br. J. Dermatol. 149: 23-29.
- Zhu, M., Lovell, K.L., Patterson, J.S., Saunders, T.L., Hughes, E.D. and Friderici, K.H. 2006. β -mannosidosis mice: a model for the human lysosomal storage disease. Hum. Mol. Genet. 15: 493-500.
- Riise Stensland, H.M., Persichetti, E., Sorriso, C., Hansen, G.M., Bibi, L., Paciotti, S., Balducci, C. and Beccari, T. 2008. Identification of two novel β -mannosidosis-associated sequence variants: biochemical analysis of β -mannosidase (MANBA) missense mutations. Mol. Genet. Metab. 94: 476-480.
- Online Mendelian Inheritance in Man, OMIM™. 2009. Johns Hopkins University, Baltimore, MD. MIM Number: 609489. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>

CHROMOSOMAL LOCATION

Genetic locus: Manba (mouse) mapping to 3 G3.

PROTOCOLS

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

PRODUCT

MANBA siRNA (m) is a pool of 3 target-specific 19-25 nt siRNAs designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10 μ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see MANBA shRNA Plasmid (m): sc-149247-SH and MANBA shRNA (m) Lentiviral Particles: sc-149247-V as alternate gene silencing products.

For independent verification of MANBA (m) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-149247A, sc-149247B and sc-149247C.

STORAGE AND RESUSPENSION

Store lyophilized siRNA duplex at -20° C with desiccant. Stable for at least one year from the date of shipment. Once resuspended, store at -20° C, avoid contact with RNAses and repeated freeze thaw cycles.

Resuspend lyophilized siRNA duplex in 330 μ l of the RNase-free water provided. Resuspension of the siRNA duplex in 330 μ l of RNase-free water makes a 10 μ M solution in a 10 μ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

MANBA siRNA (m) is recommended for the inhibition of MANBA expression in mouse cells.

SUPPORT REAGENTS

For optimal siRNA transfection efficiency, Santa Cruz Biotechnology's siRNA Transfection Reagent: sc-29528 (0.3 ml), siRNA Transfection Medium: sc-36868 (20 ml) and siRNA Dilution Buffer: sc-29527 (1.5 ml) are recommended. Control siRNAs or Fluorescein Conjugated Control siRNAs are available as 10 μ M in 66 μ l. Each contain a scrambled sequence that will not lead to the specific degradation of any known cellular mRNA. Fluorescein Conjugated Control siRNAs include: sc-36869, sc-44239, sc-44240 and sc-44241. Control siRNAs include: sc-37007, sc-44230, sc-44231, sc-44232, sc-44233, sc-44234, sc-44235, sc-44236, sc-44237 and sc-44238.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor MANBA gene expression knockdown using RT-PCR Primer: MANBA (m)-PR: sc-149247-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

RESEARCH USE

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