

BUP-1 siRNA (h): sc-62028

BACKGROUND

BUP-1 (β -ureidopropionase), also known as β -alanine synthase or N-carbamyl- β -alanine amidohydrolase, belongs to the BUP subfamily within the CN hydrolase family. BUP-1 is found in liver and kidney, localizing to the cytoplasm, and contains one CN hydrolase domain. BUP-1 catalyzes the third and last step in the degradation of thymine and uracil, the hydrolysis of N-carbamyl- β -aminoisobutyric acid (or N-carbamyl- β -alanine) to β -aminoisobutyric acid (or β -alanine), ammonia and CO₂. Deficiency in BUP-1 leads to elevated levels of N-carbamyl- β -aminoisobutyric acid and N-carbamyl- β -alanine in plasma, cerebrospinal fluid and urine, which may result in abnormal neurological activity.

REFERENCES

1. Vreken, P., et al. 1999. cDNA cloning, genomic structure and chromosomal localization of the human BUP-1 gene encoding β -ureidopropionase. *Biochim. Biophys. Acta* 1447: 251-257.
2. Moolenaar, S.H., et al. 2001. β -Ureidopropionase deficiency: a novel inborn error of metabolism discovered using NMR spectroscopy on urine. *Magn. Reson. Med.* 46: 1014-1017.
3. Sakamoto, T., et al. 2001. Expression and properties of human liver β -ureidopropionase. *J. Nutr. Sci. Vitaminol.* 47: 132-138.
4. van Kuilenburg, A.B., et al. 2002. Confirmation of the enzyme defect in the first case of β -ureidopropionase deficiency. β -alanine deficiency. *Adv. Exp. Med. Biol.* 486: 243-246.
5. van Kuilenburg, A.B., et al. 2004. β -ureidopropionase deficiency: an inborn error of pyrimidine degradation associated with neurological abnormalities. *Hum. Mol. Genet.* 13: 2793-2801.
6. Assmann, B., et al. 2006. Clinical findings and a therapeutic trial in the first patient with β -ureidopropionase deficiency. *Neuropediatrics* 37: 20-25.
7. van Kuilenburg, A.B., et al. 2006. Genetic analysis of the first 4 patients with β -ureidopropionase deficiency. *Nucleosides Nucleotides Nucleic Acids* 25: 1093-1098.

CHROMOSOMAL LOCATION

Genetic locus: UPB1 (human) mapping to 22q11.23.

PRODUCT

BUP-1 siRNA (h) 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 BUP-1 shRNA Plasmid (h): sc-62028-SH and BUP-1 shRNA (h) Lentiviral Particles: sc-62028-V as alternate gene silencing products.

For independent verification of BUP-1 (h) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-62028A, sc-62028B and sc-62028C.

RESEARCH USE

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

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

BUP-1 siRNA (h) is recommended for the inhibition of BUP-1 expression in human 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.

GENE EXPRESSION MONITORING

BUP-1 (F-11): sc-374066 is recommended as a control antibody for monitoring of BUP-1 gene expression knockdown by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) or immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ 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. 2) Immunofluorescence: use m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor BUP-1 gene expression knockdown using RT-PCR Primer: BUP-1 (h)-PR: sc-62028-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

PROTOCOLS

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