



# BUP-1 siRNA (m): sc-62029

## BACKGROUND

BUP-1 ( $\beta$ -ureidopropionase), also known as  $\beta$ -alanine synthase or N-carbamyl- $\beta$ -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- $\beta$ -aminoisobutyric acid (or N-carbamyl- $\beta$ -alanine) to  $\beta$ -aminoisobutyric acid (or  $\beta$ -alanine), ammonia and  $\text{CO}_2$ . Deficiency in BUP-1 leads to elevated levels of N-carbamyl- $\beta$ -aminoisobutyric acid and N-carbamyl- $\beta$ -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  $\beta$ -ureidopropionase. *Biochim. Biophys. Acta* 1447: 251-257.
2. Moolenaar, S.H., et al. 2001.  $\beta$ -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  $\beta$ -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  $\beta$ -ureidopropionase deficiency.  $\beta$ -alanine deficiency. *Adv. Exp. Med. Biol.* 486: 243-246.
5. van Kuilenburg, A.B., et al. 2004.  $\beta$ -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  $\beta$ -ureidopropionase deficiency. *Neuropediatrics* 37: 20-25.
7. van Kuilenburg, A.B., et al. 2006. Genetic analysis of the first 4 patients with  $\beta$ -ureidopropionase deficiency. *Nucleosides Nucleotides Nucleic Acids* 25: 1093-1098.

## CHROMOSOMAL LOCATION

Genetic locus: Upb1 (mouse) mapping to 10 C1.

## PRODUCT

BUP-1 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  $\mu\text{M}$  solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see BUP-1 shRNA Plasmid (m): sc-62029-SH and BUP-1 shRNA (m) Lentiviral Particles: sc-62029-V as alternate gene silencing products.

For independent verification of BUP-1 (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-62029A, sc-62029B and sc-62029C.

## RESEARCH USE

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

## STORAGE AND RESUSPENSION

Store lyophilized siRNA duplex at  $-20^\circ\text{C}$  with desiccant. Stable for at least one year from the date of shipment. Once resuspended, store at  $-20^\circ\text{C}$ , avoid contact with RNases and repeated freeze thaw cycles.

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

## APPLICATIONS

BUP-1 siRNA (m) is recommended for the inhibition of BUP-1 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  $\mu\text{M}$  in 66  $\mu\text{l}$ . 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 $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  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 $\kappa$  BP-FITC: sc-516140 or m-IgG $\kappa$  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 (m)-PR: sc-62029-PR (20  $\mu\text{l}$ ). Annealing temperature for the primers should be  $55-60^\circ\text{C}$  and the extension temperature should be  $68-72^\circ\text{C}$ .

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) for detailed protocols and support products.