

## BUP-1 (A-3): sc-374345

### 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 CO<sub>2</sub>. 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. van Kuilenburg, A.B., et al. 2000. Confirmation of the enzyme defect in the first case of  $\beta$ -ureidopropionase deficiency.  $\beta$ -alanine deficiency. *Adv. Exp. Med. Biol.* 486: 243-246.
3. 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.
4. Sakamoto, T., et al. 2001. Expression and properties of human liver  $\beta$ -ureidopropionase. *J. Nutr. Sci. Vitaminol.* 47: 132-138.
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 (human) mapping to 22q11.23; Upb1 (mouse) mapping to 10 C1.

### SOURCE

BUP-1 (A-3) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 277-311 within an internal region of BUP-1 of human origin.

### PRODUCT

Each vial contains 200  $\mu$ g IgG<sub>1</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-374345 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

### APPLICATIONS

BUP-1 (A-3) is recommended for detection of BUP-1 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2  $\mu$ g per 100-500  $\mu$ g of total protein (1 ml of cell lysate)], 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).

Suitable for use as control antibody for BUP-1 siRNA (h): sc-62028, BUP-1 siRNA (m): sc-62029, BUP-1 shRNA Plasmid (h): sc-62028-SH, BUP-1 shRNA Plasmid (m): sc-62029-SH, BUP-1 shRNA (h) Lentiviral Particles: sc-62028-V and BUP-1 shRNA (m) Lentiviral Particles: sc-62029-V.

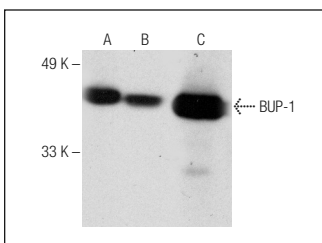
Molecular Weight of BUP-1: 43 kDa.

Positive Controls: human liver extract: sc-363766, rat liver extract: sc-2395 or rat kidney extract: sc-2394.

### RECOMMENDED SUPPORT REAGENTS

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<sup>™</sup> Molecular Weight Standards: sc-2035, UltraCruz<sup>®</sup> Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) 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<sup>®</sup> Mounting Medium: sc-24941 or UltraCruz<sup>®</sup> Hard-set Mounting Medium: sc-359850.

### DATA



BUP-1 (A-3): sc-374345. Western blot analysis of BUP-1 expression in rat liver (A) rat kidney (B) and human liver (C) tissue extracts.

### 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](http://www.scbt.com) for detailed protocols and support products.