

# BUP-1 (V-17): sc-66609

## 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. Assmann, B.E., et al. 2006.  $\beta$ -ureidopropionase deficiency presenting with febrile status epilepticus. *Epilepsia* 47: 215-217.

## CHROMOSOMAL LOCATION

Genetic locus: UPB1 (human) mapping to 22q11.23; Upb1 (mouse) mapping to 10 C1.

## SOURCE

BUP-1 (V-17) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of BUP-1 of human origin.

## PRODUCT

Each vial contains 200  $\mu\text{g}$  IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-66609 P, (100  $\mu\text{g}$  peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## STORAGE

Store at 4° C, \*\*DO NOT FREEZE\*\*. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

## APPLICATIONS

BUP-1 (V-17) is recommended for detection of BUP-1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2  $\mu\text{g}$  per 100-500  $\mu\text{g}$  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).

BUP-1 (V-17) is also recommended for detection of BUP-1 in additional species, including equine, canine, bovine and porcine.

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

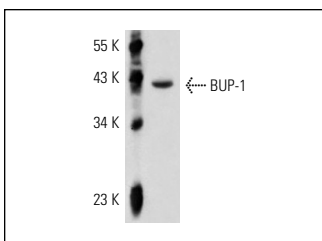
Molecular Weight of BUP-1: 43 kDa.

Positive Controls: rat liver extract: sc-2395 or mouse liver extract: sc-2256.

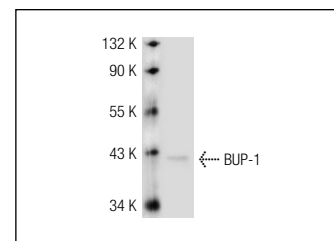
## RECOMMENDED SECONDARY REAGENTS

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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) 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.

## DATA



BUP-1 (V-17): sc-66609. Western blot analysis of BUP-1 expression in rat liver tissue extract.



BUP-1 (V-17): sc-66609. Western blot analysis of BUP-1 expression in mouse liver tissue extract.

## 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) or our catalog for detailed protocols and support products.