

BUP-1 (B-5): sc-393221

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 CO_2 . 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.
8. Assmann, B.E., et al. 2006. β -ureidopropionase deficiency presenting with febrile status epilepticus. *Epilepsia* 47: 215-217.

CHROMOSOMAL LOCATION

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

SOURCE

BUP-1 (B-5) is a mouse monoclonal antibody raised against amino acids 164-384 mapping at the C-terminus of BUP-1 of human origin.

PRODUCT

Each vial contains 200 μg IgG $_1$ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

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 (B-5) is recommended for detection of BUP-1 of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μg per 100-500 μg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (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 shRNA Plasmid (h): sc-62028-SH and BUP-1 shRNA (h) Lentiviral Particles: sc-62028-V.

Molecular Weight of BUP-1: 43 kDa.

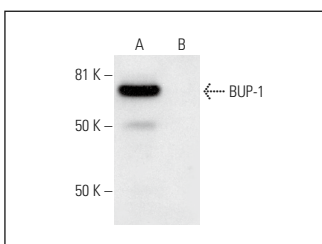
Positive Controls: human liver extract: sc-363766.

RECOMMENDED SUPPORT REAGENTS

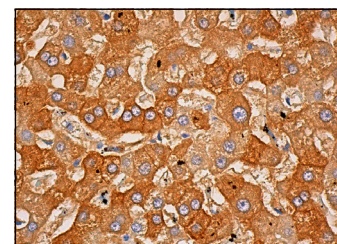
To ensure optimal results, the following support reagents are recommended:

- 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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).
- 3) 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.
- 4) Immunohistochemistry: use m-IgG κ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



BUP-1 (B-5): sc-393221. Western blot analysis of BUP-1 expression in human liver (A) and rat liver (B) tissue extracts. Note lack of reactivity with rat BUP-1 in lane B.



BUP-1 (B-5): sc-393221. Immunoperoxidase staining of formalin fixed, paraffin-embedded human liver tissue showing cytoplasmic and membrane staining of hepatocytes.

RESEARCH USE

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

PROTOCOLS

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