SANTA CRUZ BIOTECHNOLOGY, INC.

BUP-1 (G-9): sc-390541

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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. Assmann, B.E., et al. 2006. β-ureidopropionase deficiency presenting with febrile status epilepticus. Epilepsia 47: 215-217.
8. 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; Upb1 (mouse) mapping to 10 C1.

SOURCE

BUP-1 (G-9) 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.

APPLICATIONS

BUP-1 (G-9) 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 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 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.

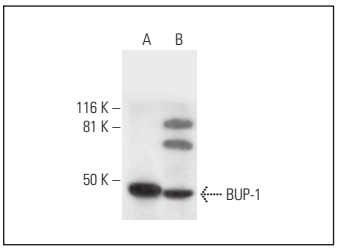
Molecular Weight of BUP-1: 43 kDa.

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

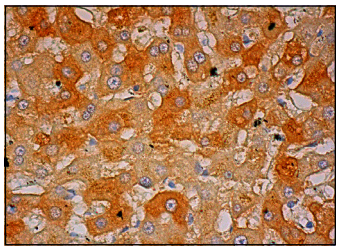
RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ 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-IgGκ BP-FITC: sc-516140 or m-IgGκ 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-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



BUP-1 (G-9): sc-390541. Western blot analysis of BUP-1 expression in human liver (**A**) and rat liver (**B**) tissue extracts.



BUP-1 (G-9): sc-390541. Immunoperoxidase staining of formalin fixed, paraffin-embedded human liver tissue showing cytoplasmic staining of hepatocytes.

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.