SANTA CRUZ BIOTECHNOLOGY, INC.

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

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. van Kuilenburg, A.B., et al. 2000. Confirmation of the enzyme defect in the first case of β-ureidopropionase deficiency. β-alanine deficiency. Adv. Exp. Med. Biol. 486: 243-246.
3. 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.
4. Sakamoto, T., et al. 2001. Expression and properties of human liver β-ureidopropionase. J. Nutr. Sci. Vitaminol. 47: 132-138.
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.

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 µg IgG_1 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 µg 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 µg per 100-500 µg 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.

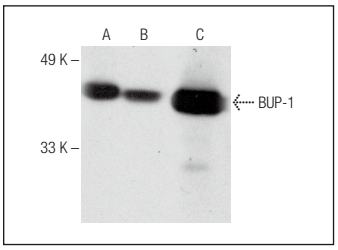
Molecular Weight of BUP-1: 43 kDa.

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-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.

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 for detailed protocols and support products.