

BUP-1 (F-11): sc-374066

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.

CHROMOSOMAL LOCATION

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

SOURCE

BUP-1 (F-11) 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_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

BUP-1 (F-11) is available conjugated to agarose (sc-374066 AC), 500 μg /0.25 ml agarose in 1 ml, for IP; to HRP (sc-374066 HRP), 200 $\mu\text{g}/\text{ml}$, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-374066 PE), fluorescein (sc-374066 FITC), Alexa Fluor[®] 488 (sc-374066 AF488), Alexa Fluor[®] 546 (sc-374066 AF546), Alexa Fluor[®] 594 (sc-374066 AF594) or Alexa Fluor[®] 647 (sc-374066 AF647), 200 $\mu\text{g}/\text{ml}$, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor[®] 680 (sc-374066 AF680) or Alexa Fluor[®] 790 (sc-374066 AF790), 200 $\mu\text{g}/\text{ml}$, for Near-Infrared (NIR) WB, IF and FCM.

Blocking peptide available for competition studies, sc-374066 P, (100 μg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

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APPLICATIONS

BUP-1 (F-11) 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.

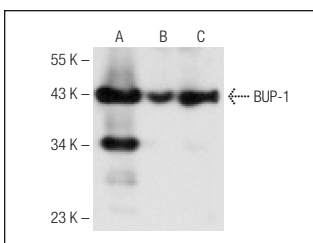
Molecular Weight of BUP-1: 43 kDa.

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

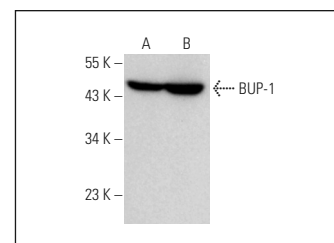
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-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.

DATA



BUP-1 (F-11): sc-374066. Western blot analysis of BUP-1 expression in human liver (A), rat liver (B) and rat kidney (C) tissue extracts.



BUP-1 (F-11): sc-374066. Western blot analysis of BUP-1 expression in rat liver (A) and mouse liver (B) 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.