

BBOX1 (E-11): sc-373774

BACKGROUND

BBOX1 (butyrobetaine γ , 2-oxoglutarate dioxygenase 1), also known as BBH, BBOX, G-BBH (γ -butyrobetaine hydroxylase) or γ -BBH, is a member of the γ -BBH/TMLD family of proteins. Localizing to the cytoplasm and predominantly expressed in kidney, BBOX1 plays a role in the biosynthesis of amines, polyamines and carnitines. More specifically, BBOX1 catalyzes the hydroxylation of γ -butyrobetaine to L-carnitine, a reaction that requires iron and ascorbate as cofactors. This is the last of five steps comprising the L-carnitine biosynthesis pathway and it is important for the proper transport of activated fatty acids across the mitochondrial membrane. Coupled to this reaction, BBOX1 simultaneously catalyzes the oxidative decarboxylation of α -ketoglutarate to succinate.

REFERENCES

- Lindstedt, G., et al. 1982. γ -butyrobetaine hydroxylase in human kidney. *Scand. J. Clin. Lab. Invest.* 42: 477-485.
- Lindstedt, S., et al. 1984. Multiple forms of γ -butyrobetaine hydroxylase (EC 1.14.11.1). *Biochem. J.* 223: 119-127.
- Vaz, F.M., et al. 1998. Carnitine biosynthesis: identification of the cDNA encoding human γ -butyrobetaine hydroxylase. *Biochem. Biophys. Res. Commun.* 250: 506-510.
- Galland, S., et al. 1998. Purification and characterization of the rat liver γ -butyrobetaine hydroxylase. *Mol. Cell. Biochem.* 178: 163-168.
- Galland, S., et al. 1999. Molecular cloning and characterization of the cDNA encoding the rat liver γ -butyrobetaine hydroxylase. *Biochim. Biophys. Acta* 1441: 85-92.
- Galland, S., et al. 2002. Thyroid hormone controls carnitine status through modifications of γ -butyrobetaine hydroxylase activity and gene expression. *Cell. Mol. Life Sci.* 59: 540-545.

CHROMOSOMAL LOCATION

Genetic locus: BBOX1 (human) mapping to 11p14.2.

SOURCE

BBOX1 (E-11) is a mouse monoclonal antibody raised against amino acids 47-346 mapping within an internal region of BBOX1 of human origin.

PRODUCT

Each vial contains 200 μ g IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

BBOX1 (E-11) is available conjugated to agarose (sc-373774 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-373774 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-373774 PE), fluorescein (sc-373774 FITC), Alexa Fluor® 488 (sc-373774 AF488), Alexa Fluor® 546 (sc-373774 AF546), Alexa Fluor® 594 (sc-373774 AF594) or Alexa Fluor® 647 (sc-373774 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-373774 AF680) or Alexa Fluor® 790 (sc-373774 AF790), 200 μ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

BBOX1 (E-11) is recommended for detection of BBOX1 of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g 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 BBOX1 siRNA (h): sc-96811, BBOX1 shRNA Plasmid (h): sc-96811-SH and BBOX1 shRNA (h) Lentiviral Particles: sc-96811-V.

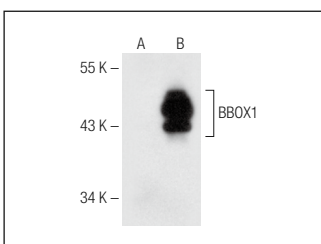
Molecular Weight of BBOX1: 45 kDa.

Positive Controls: BBOX1 (h): 293T Lysate: sc-171548.

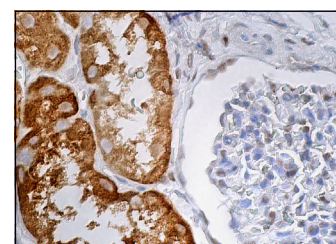
RECOMMENDED SUPPORT REAGENTS

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



BBOX1 (E-11): sc-373774. Western blot analysis of BBOX1 expression in non-transfected: sc-117752 (A) and human BBOX1 transfected: sc-171548 (B) 293T whole cell lysates.



BBOX1 (E-11): sc-373774. Immunoperoxidase staining of formalin fixed, paraffin-embedded human kidney tissue showing cytoplasmic staining of cells in tubules.

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.