

BBOX1 (H-300): sc-134963

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. and Nordin, I. 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; Bbox1 (mouse) mapping to 2 E3.

SOURCE

BBOX1 (H-300) is a rabbit polyclonal antibody raised against amino acids 47-346 mapping within an internal region of BBOX1 of human origin.

PRODUCT

Each vial contains 200 μ g IgG 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.

PROTOCOLS

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

APPLICATIONS

BBOX1 (H-300) is recommended for detection of BBOX1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

BBOX1 (H-300) is also recommended for detection of BBOX1 in additional species, including equine, canine and bovine.

Suitable for use as control antibody for BBOX1 siRNA (h): sc-96811, BBOX1 siRNA (m): sc-141481, BBOX1 shRNA Plasmid (h): sc-96811-SH, BBOX1 shRNA Plasmid (m): sc-141481-SH, BBOX1 shRNA (h) Lentiviral Particles: sc-96811-V and BBOX1 shRNA (m) Lentiviral Particles: sc-141481-V.

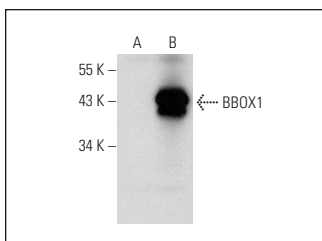
Molecular Weight of BBOX1: 45 kDa.

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

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



BBOX1 (H-300): sc-134963. Western blot analysis of BBOX1 expression in non-transfected: sc-117752 (A) and human BBOX1 transfected: sc-171548 (B) 293T whole cell lysates.

RESEARCH USE

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



Try **BBOX1 (E-11): sc-373774**, our highly recommended monoclonal alternative to BBOX1 (H-300).