

BBOX1 (h): 293T Lysate: sc-171548

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

1. Lindstedt, G., et al. 1982. γ -butyrobetaine hydroxylase in human kidney. *Scand. J. Clin. Lab. Invest.* 42: 477-485.
2. Lindstedt, S. and Nordin, I. 1984. Multiple forms of γ -butyrobetaine hydroxylase (EC 1.14.11.1). *Biochem. J.* 223: 119-127.
3. Vaz, F.M., et al. 1998. Carnitine biosynthesis: identification of the cDNA encoding human γ -butyrobetaine hydroxylase. *Biochem. Biophys. Res. Commun.* 250: 506-510.
4. Galland, S., et al. 1998. Purification and characterization of the rat liver γ -butyrobetaine hydroxylase. *Mol. Cell. Biochem.* 178: 163-168.
5. 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.
6. 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.
7. Verheyen, G.R., et al. 2004. Microarray analysis of the effect of diesel exhaust particles on *in vitro* cultured macrophages. *Toxicol. In Vitro* 18: 377-391.

CHROMOSOMAL LOCATION

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

PRODUCT

BBOX1 (h): 293T Lysate represents a lysate of human BBOX1 transfected 293T cells and is provided as 100 μ g protein in 200 μ l SDS-PAGE buffer.

APPLICATIONS

BBOX1 (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive BBOX1 antibodies. Recommended use: 10-20 μ l per lane.

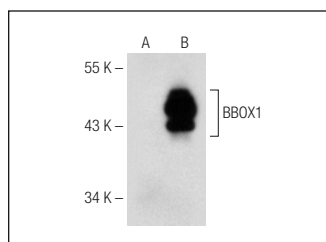
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

BBOX1 (E-11): sc-373774 is recommended as a positive control antibody for Western Blot analysis of enhanced human BBOX1 expression in BBOX1 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

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.

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.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. 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.