

BCKDHB (H-300): sc-366071

BACKGROUND

BCKDHB (branched chain keto acid dehydrogenase E1, β polypeptide), also known as 2-oxoisovalerate dehydrogenase subunit β mitochondrial or E1B, is a 392 amino acid mitochondrial matrix protein and component of branched-chain keto acid dehydrogenase, a multienzyme complex involved in the catabolism of branched-chain amino acids. Existing as a heterodimer, BCKDHB is encoded by a gene mapping to human chromosome 6q14.1, whose defects are the cause of an autosomal recessive disorder known as maple syrup urine disease type 1B (MSUD1B). Characterized by urine with maple syrup odor, patients with maple syrup urine disease may suffer severe neurological damage, mental retardation and feeding problems.

REFERENCES

1. Chuang, J.L., et al. 1990. Molecular cloning of the mature E1b- β subunit of human branched-chain α -keto acid dehydrogenase complex. *FEBS Lett.* 262: 305-309.
2. Zneimer, S.M., et al. 1991. Regional assignment of two genes of the human branched-chain α -keto acid dehydrogenase complex: the E1 β gene (BCKDHB) to chromosome 6p21-22 and the E2 gene (DBT) to chromosome 1p31. *Genomics* 10: 740-747.

CHROMOSOMAL LOCATION

Genetic locus: BCKDHB (human) mapping to 6q14.1; Bckdhd (mouse) mapping to 9 E2.

SOURCE

BCKDHB (H-300) is a rabbit polyclonal antibody raised against amino acids 1-300 mapping at the N-terminus of BCKDHB 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.

APPLICATIONS

BCKDHB (H-300) is recommended for detection of BCKDHB 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).

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

Suitable for use as control antibody for BCKDHB siRNA (h): sc-95231, BCKDHB siRNA (m): sc-141668, BCKDHB shRNA Plasmid (h): sc-95231-SH, BCKDHB shRNA Plasmid (m): sc-141668-SH, BCKDHB shRNA (h) Lentiviral Particles: sc-95231-V, and BCKDHB shRNA (m) Lentiviral Particles: sc-141668-V.

Molecular Weight (predicted) of BCKDHB: 43 kDa.

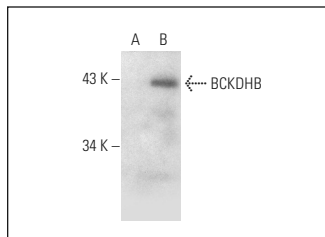
Molecular Weight (observed) of BCKDHB: 43-55 kDa.

Positive Controls: BCKDHB (h): 293T Lysate: sc-115538.

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



BCKDHB (H-300): sc-366071. Western blot analysis of BCKDHB expression in non-transfected: sc-117752 (A) and human BCKDHB transfected: sc-115538 (B) 293T whole cell lysates.

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 or our catalog for detailed protocols and support products.



Try **BCKDHB (H-6): sc-374630**, our highly recommended monoclonal alternative to BCKDHB (H-300).