SANTA CRUZ BIOTECHNOLOGY, INC.

BCKDHB (H-6): sc-374630

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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 IB (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.
3. Patel, M.S. and Harris, R.A. 1995. Mammalian α-keto acid dehydrogenase complexes: gene regulation and genetic defects. FASEB J. 9: 1164-1172.

CHROMOSOMAL LOCATION

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

SOURCE

BCKDHB (H-6) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 67-97 within an internal region of BCKDHB of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

BCKDHB (H-6) is available conjugated to agarose (sc-374630 AC), 500 µg/ 0.25 ml agarose in 1 ml, for IP; to HRP (sc-374630 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-374630 PE), fluorescein (sc-374630 FITC), Alexa Fluor® 488 (sc-374630 AF488), Alexa Fluor® 546 (sc-374630 AF546), Alexa Fluor® 594 (sc-374630 AF594) or Alexa Fluor® 647 (sc-374630 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-374630 AF680) or Alexa Fluor® 790 (sc-374630 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

Blocking peptide available for competition studies, sc-374630 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

APPLICATIONS

BCKDHB (H-6) 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 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).

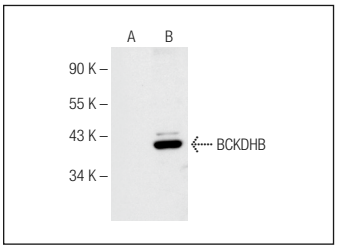
Suitable for use as control antibody for BCKDHB siRNA (h): sc-95231, BCKDHB siRNA (m): sc-141668, BCKDHB siRNA (r): sc-270287, BCKDHB shRNA Plasmid (h): sc-95231-SH, BCKDHB shRNA Plasmid (m): sc-141668-SH, BCKDHB shRNA Plasmid (r): sc-270287-SH, BCKDHB shRNA (h) Lentiviral Particles: sc-95231-V, BCKDHB shRNA (m) Lentiviral Particles: sc-141668-V and BCKDHB shRNA (r) Lentiviral Particles: sc-270287-V.

Molecular Weight (predicted) of BCKDHB: 43 kDa.

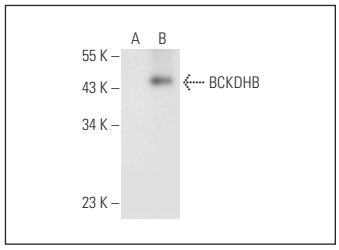
Molecular Weight (observed) of BCKDHB: 43-55 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, mouse liver extract: sc-2256 or BCKDHB (h): 293T Lysate: sc-115538.

DATA



BCKDHB (H-6): sc-374630. Western blot analysis of BCKDHB expression in non-transfected (**A**) and human BCKDHB transfected (**B**) HEK293T whole cell lysates.



BCKDHB (H-6): sc-374630. Western blot analysis of BCKDHB expression in non-transfected: sc-117752 (**A**) and human BCKDHB transfected: sc-115538 (**B**) 293T whole cell lysates.

SELECT PRODUCT CITATIONS

1. Yue, S.J., et al. 2019. Berberine alleviates Insulin resistance by reducing peripheral branched-chain amino acids. Am. J. Physiol. Endocrinol. Metab. 316: E73-E85.
2. Tian, N., et al. 2020. Transketolase deficiency in adipose tissues protects mice from diet-induced obesity by promoting lipolysis. Diabetes 69: 1355-1367.
3. Biswas, D., et al. 2020. Adverse outcomes in obese cardiac surgery patients correlates with altered branched-chain amino acid catabolism in adipose tissue and heart. Front. Endocrinol. 11: 534.

RESEARCH USE

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