SANTA CRUZ BIOTECHNOLOGY, INC.

BCKDHB (E-15): sc-160972

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., Cox, R.P. and Chuang, D.T. 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., Lau, K.S., Eddy, R.L., Shows, T.B., Chuang, J.L., Chuang, D.T. and Cox, R.P. 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.
4. Chuang, J.L., Cox and R.P., Chuang, D.T. 1996. Maple syrup urine disease: the E1β gene of human branched-chain α-ketoacid dehydrogenase complex has 11 rather than 10 exons, and the 3' UTR in one of the two E1β mRNAs arises from intronic sequences. Am. J. Hum. Genet. 58: 1373-1377.
5. Chuang, D.T. 1998. Maple syrup urine disease: it has come a long way. J. Pediatr. 132: S17-S23.

CHROMOSOMAL LOCATION

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

SOURCE

BCKDHB (E-15) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of BCKDHB of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-160972 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

BCKDHB (E-15) 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), 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 (E-15) is also recommended for detection of BCKDHB in additional species, including equine, canine, bovine, porcine and avian.

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.

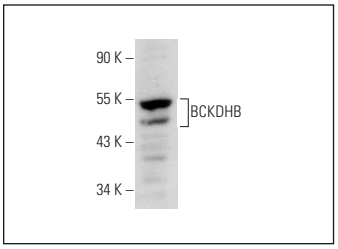
Molecular Weight (observed) of BCKDHB: 43-55 kDa.

Positive Controls: K-562 whole cell lysate: sc-2203.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (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) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



BCKDHB (E-15): sc-160972. Western blot analysis of BCKDHB expression in K-562 whole cell lysate.

RESEARCH USE

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

MONOS Satisfaction Guaranteed

Try **BCKDHB (H-6): sc-374630**, our highly recommended monoclonal alternative to BCKDHB (E-15).