SANTA CRUZ BIOTECHNOLOGY, INC.

BCKDE1A (H-5): sc-271538

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

BCKDE1A (branched-chain α-keto acid dehydrogenase E1 component α chain), also known as BCKDHA or 2-oxoisovalerate dehydrogenase subunit α, is part of the inner mitochondrial membrane complex involved in the catabolism of the branched-chain amino acids. This complex consists of multiple copies of three catalytic components: BCKDE1, DBT and DLD. It is responsible for catalyzing the conversion of α-keto acids to acyl-CoA and CO_2. BCKDE1A is the α chain component of BCKDE1. BCKDE1 is heterotetrameric, consisting of two α chains and two β chains. A mutation in BCKDE1A can result in a deficiency of the properly assembled BCKDH complex. A deficiency of this enzyme leads to an accumulation of branched-chain amino acids in the blood and urine. This metabolic disorder is called type IA maple syrup urine disease (MSUD).

CHROMOSOMAL LOCATION

Genetic locus: BCKDHA (human) mapping to 19q13.2; Bckdha (mouse) mapping to 7 A3.

SOURCE

BCKDE1A (H-5) is a mouse monoclonal antibody raised against amino acids 47-346 mapping near the N-terminus of BCKDE1A of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin. Also available as TransCruz reagent for Gel Supershift and ChIP applications, sc-271538 X, 200 µg/0.1 ml.

BCKDE1A (H-5) is available conjugated to agarose (sc-271538 AC), 500 µg/ 0.25 ml agarose in 1 ml, for IP; to HRP (sc-271538 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-271538 PE), fluorescein (sc-271538 FITC), Alexa Fluor® 488 (sc-271538 AF488), Alexa Fluor® 546 (sc-271538 AF546), Alexa Fluor® 594 (sc-271538 AF594) or Alexa Fluor® 647 (sc-271538 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-271538 AF680) or Alexa Fluor® 790 (sc-271538 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

BCKDE1A (H-5) is recommended for detection of BCKDE1A of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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), immunohistochemistry (including paraffin-embedded sections) (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 BCKDE1A siRNA (h): sc-105117, BCKDE1A siRNA (m): sc-77386, BCKDE1A shRNA Plasmid (h): sc-105117-SH, BCKDE1A shRNA Plasmid (m): sc-77386-SH, BCKDE1A shRNA (h) Lentiviral Particles: sc-105117-V and BCKDE1A shRNA (m) Lentiviral Particles: sc-77386-V.

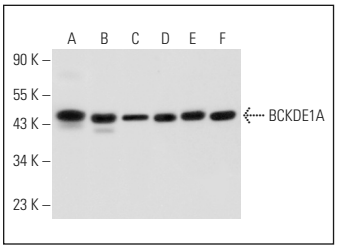
BCKDE1A (H-5) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

Molecular Weight of BCKDE1A: 50 kDa.

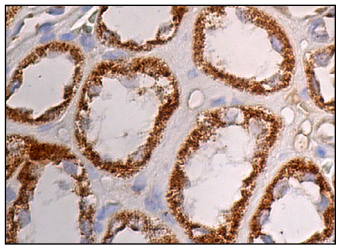
STORAGE

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

DATA



BCKDE1A (H-5): sc-271538. Western blot analysis of BCKDE1A expression in human kidney tissue extract (**A**) and Jurkat (**B**), U-251-MG (**C**), Hep G2 (**D**), K-562 (**E**) and NIH/3T3 (**F**) whole cell lysates.



BCKDE1A (H-5): sc-271538. Immunoperoxidase staining of formalin fixed, paraffin-embedded human kidney tissue showing cytoplasmic staining of cells in tubules.

SELECT PRODUCT CITATIONS

1. Yoneshiro, T., et al. 2019. BCAA catabolism in brown fat controls energy homeostasis through SLC25A44. Nature 572: 614-619.
2. Basit, F. and de Vries, I.J.M. 2019. Dendritic cells require PINK1-mediated phosphorylation of BCKDE1α to promote fatty acid oxidation for immune function. Front. Immunol. 10: 2386.
3. Lee, J.H., et al. 2019. Branched-chain amino acids sustain pancreatic cancer growth by regulating lipid metabolism. Exp. Mol. Med. 51: 1-11.
4. Tian, N., et al. 2020. Transketolase deficiency in adipose tissues protects mice from diet-induced obesity by promoting lipolysis. Diabetes 69: 1355-1367.
5. Koike, S., et al. 2020. An increase in liver polyamine concentration contributes to the tryptophan-induced acute stimulation of rat hepatic protein synthesis. Nutrients 12: 2665.
6. Kim, J.H., et al. 2021. BIX01294 inhibits EGFR signaling in EGFR-mutant lung adenocarcinoma cells through a BCKDHA-mediated reduction in the EGFR level. Exp. Mol. Med. 53: 1877-1887.
7. He, Q.Z., et al. 2022. 3,6-dichlorobenzo[b]thiophene-2-carboxylic acid alleviates ulcerative colitis by suppressing mammalian target of rapamycin complex 1 activation and regulating intestinal microbiota. World J. Gastroenterol. 28: 6522-6536.

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.