

BCKDE1A (H-5): sc-271538

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 CO₂. 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 μ g 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 μ g/0.1 ml.

BCKDE1A (H-5) is available conjugated to agarose (sc-271538 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-271538 HRP), 200 μ g/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 μ g/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 μ g/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 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), 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.

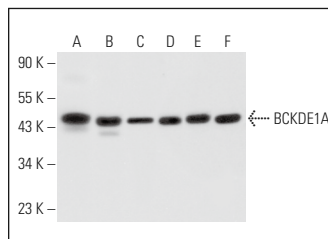
BCKDE1A (H-5) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

Molecular Weight of BCKDE1A: 50 kDa.

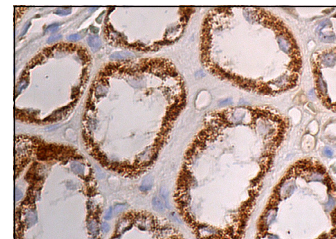
STORAGE

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

- Yoneshiro, T., et al. 2019. BCAA catabolism in brown fat controls energy homeostasis through SLC25A44. *Nature* 572: 614-619.
- Basit, F. and de Vries, I.J.M. 2019. Dendritic cells require PINK1-mediated phosphorylation of BCKDE1 α to promote fatty acid oxidation for immune function. *Front. Immunol.* 10: 2386.
- Lee, J.H., et al. 2019. Branched-chain amino acids sustain pancreatic cancer growth by regulating lipid metabolism. *Exp. Mol. Med.* 51: 1-11.
- Tian, N., et al. 2020. Transketolase deficiency in adipose tissues protects mice from diet-induced obesity by promoting lipolysis. *Diabetes* 69: 1355-1367.
- 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.
- 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.
- 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.

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