

BCKDE1A (H-300): sc-67200

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-300) is a rabbit polyclonal 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 in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

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

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.

APPLICATIONS

BCKDE1A (H-300) is recommended for detection of BCKDE1A 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).

BCKDE1A (H-300) is also recommended for detection of BCKDE1A in additional species, including equine, canine, bovine, porcine and avian.

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.

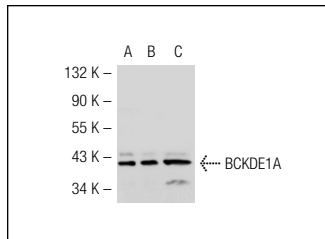
Molecular Weight of BCKDE1A: 50 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200 or BCKDE1A (m): 293T Lysate: sc-126497.

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



BCKDE1A (H-300): sc-67200. Western blot analysis of BCKDE1A expression in non-transfected 293T: sc-117752 (A), mouse BCKDE1A transfected 293T: sc-126497 (B) and HeLa (C) whole cell lysates.

RESEARCH USE

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

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Try **BCKDE1A (H-5): sc-271538**, our highly recommended monoclonal alternative to BCKDE1A (H-300).