

# BCKDE1A (G-18): sc-51471

## BACKGROUND

BCKDE1A (branched-chain  $\alpha$ -keto acid dehydrogenase E1 component  $\alpha$  chain), also known as BCKDHA or 2-oxoisovalerate dehydrogenase subunit  $\alpha$ , 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  $\alpha$ -keto acids to acyl-CoA and CO<sub>2</sub>. BCKDE1A is the  $\alpha$  chain component of BCKDE1. BCKDE1 is heterotetrameric, consisting of two  $\alpha$  chains and two  $\beta$  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 (G-18) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of BCKDE1A of human origin.

## PRODUCT

Each vial contains 200  $\mu$ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-51471 P, (100  $\mu$ g 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

BCKDE1A (G-18) is recommended for detection of precursor and mature BCKDE1A 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), 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).

BCKDE1A (G-18) is also recommended for detection of BCKDE1A precursor and mature 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.

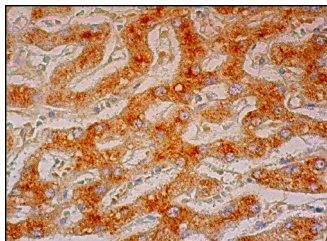
Molecular Weight of BCKDE1A: 50 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200.

## 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. 3) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

## DATA



BCKDE1A (G-18): sc-51471. Immunoperoxidase staining of formalin fixed, paraffin-embedded human liver tissue showing cytoplasmic staining of hepatocytes.

## RESEARCH USE

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

## PROTOCOLS

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Try **BCKDE1A (H-5): sc-271538**, our highly recommended monoclonal alternative to BCKDE1A (G-18).