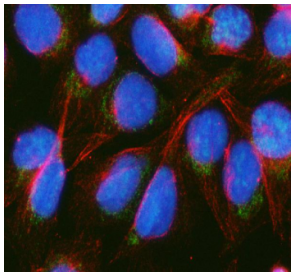
BCKDH E2 Antibody / Dihydrolipoamide branched chain transacylase E2 / DBT (FY13381)

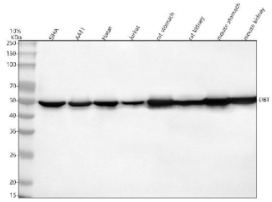
Catalog No.	Formulation	Size
FY13381	Adding 0.2 ml of distilled water will yield a concentration of 500 ug/ml	100 ug

Bulk quote request

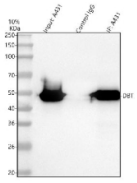
Availability	1-2 days
Species Reactivity	Human, Mouse, Rat
Format	Lyophilized
Host	Rabbit
Clonality	Polyclonal (rabbit origin)
Isotype	Rabbit IgG
Purity	Immunogen affinity purified
Buffer	Each vial contains 4 mg Trehalose, 0.9 mg NaCl, 0.2 mg Na2HPO4.
UniProt	P11182
Localization	Mitochondria
Applications	Western Blot : 0.25-0.5ug/ml Immunocytochemistry/Immunofluorescence : 5ug/ml Immunoprecipitation : 2-4ug/500ug of lysate Flow Cytometry : 1-3ug/million cells ELISA : 0.1-0.5ug/ml
Limitations	This BCKDH E2 antibody is available for research use only.



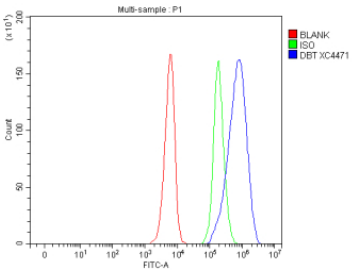
Immunofluorescent staining of BCKDH E2/DBT using anti-BCKDH E2 antibody (green) and anti-Beta Tubulin antibody (red). BCKDH E2/DBT was detected in an immunocytochemical section of U2OS cells. Enzyme antigen retrieval was performed using IHC enzyme antigen retrieval reagent for 15 mins. The cells were blocked with 10% goat serum. And then incubated with 5 ug/ml rabbit anti-BCKDH E2 antibody and mouse anti-Beta Tubulin antibody overnight at 4oC. DyLight 488 Conjugated Goat Anti-Rabbit IgG and DyLight 550 Conjugated Goat Anti-Mouse IgG were used as secondary antibody at 1:500 dilution and incubated for 30 minutes at 37oC. The section was counterstained with DAPI nuclear stain (blue). Visualize using a fluorescence microscope and filter sets appropriate for the label used.



Western blot analysis of BCKDH E2/DBT using anti-BCKDH E2 antibody. Lane 1: human SiHa whole cell lysates, Lane 2: human whole cell lysates, Lane 3: human Hacat whole cell lysates, Lane 4: human Jurkat whole cell lysates, Lane 5: rat stomach tissue lysates, Lane 6: rat kidney tissue lysates, Lane 7: mouse stomach tissue lysates, Lane 8: mouse kidney tissue lysates. After electrophoresis, proteins were transferred to a nitrocellulose membrane at 150 mA for 50-90 minutes. Blocked the membrane with 5% non-fat milk/TBS for 1.5 hour at RT. The membrane was incubated with rabbit anti-BCKDH E2 antibody at 0.5 ug/ml overnight at 4oC, then washed with TBS-0.1%Tween 3 times with 5 minutes each and probed with a goat anti-rabbit IgG-HRP secondary antibody at a dilution of 1:5000 for 1.5 hour at RT. The signal was developed using enhanced chemiluminescent. BCKDH E2 (DBT) antibody detects a single strong band just below the 50 kDa marker in human cell and tissue lysates, slightly faster than the 53.5 kDa predicted mass but consistent with the processed 52 kDa mitochondrial autoantigen form of DBT reported in the literature and typical SDS-PAGE migration behavior of mitochondrial enzymes.



Immunoprecipitating BCKDH E2/DBT in whole cell lysate. Western blot analysis of BCKDH E2/DBT using anti-BCKDH E2 antibody. Lane 1: whole cell lysates (30ug), Lane 2: Rabbit control IgG instead of anti-BCKDH E2 antibody in whole cell lysate, Lane 3: anti-BCKDH E2 antibody (2ug) + whole cell lysate (500ug). After electrophoresis, proteins were transferred to a membrane. Then the membrane was incubated with rabbit anti-BCKDH E2 antibody at a dilution of 0.5 ug/ml and probed with a mouse anti-rabbit IgG-HRP secondary antibody (Light Chain). The signal is developed using ECL Plus Western Blotting Substrate. BCKDH E2 (DBT) antibody detects a single strong band just below the 50 kDa marker in human cell and tissue lysates, slightly faster than the 53.5 kDa predicted mass but consistent with the processed 52 kDa mitochondrial autoantigen form of DBT reported in the literature and typical SDS-PAGE migration behavior of mitochondrial enzymes.



Flow Cytometry analysis of human SiHa cells using anti-BCKDH E2 antibody. Overlay histogram showing SiHa cells stained with (Blue line). To facilitate intracellular staining, cells were fixed with 4% paraformaldehyde and permeabilized with permeabilization buffer. The cells were blocked with 10% normal goat serum. And then incubated with rabbit anti-BCKDH E2 antibody (1 ug/million cells) for 30 min at 20oC. DyLight 488 conjugated goat anti-rabbit IgG (5-10 ug/million cells) was used as secondary antibody for 30 minutes at 20oC. Isotype control antibody (Green line) was rabbit IgG (1 ug/million cells) used under the same conditions. Unlabelled sample without incubation with primary antibody and secondary antibody (Red line) was used as a blank control.

Description

BCKDH E2 antibody detects Dihydrolipoamide branched chain transacylase E2, the core component of the branched-chain alpha-keto acid dehydrogenase (BCKDH) complex encoded by the DBT gene on chromosome 1p31.3. This mitochondrial enzyme catalyzes a critical step in the oxidative decarboxylation of branched-chain amino acids (BCAAs)-leucine, isoleucine, and valine-linking amino acid catabolism to energy metabolism. The BCKDH E2 subunit serves as the central scaffold of the multienzyme complex, coordinating interactions with E1 (branched-chain alpha-keto acid decarboxylase) and E3 (dihydrolipoamide dehydrogenase) components to facilitate efficient substrate channeling within mitochondria.

BCKDH E2 belongs to the 2-oxo acid dehydrogenase family and forms a 24-mer cubic core structure that anchors multiple copies of the peripheral E1 and E3 enzymes. Each E2 subunit contains lipoyl domains that shuttle reaction intermediates between catalytic sites, an inner-core acyltransferase domain, and flexible linker regions that enable conformational movement during catalysis. The enzyme's lipoyl-lysine cofactors play a pivotal role in transferring acyl groups to CoA, producing acyl-CoA derivatives for entry into the tricarboxylic acid (TCA) cycle. Co-localization studies

confirm mitochondrial matrix localization, consistent with its role in oxidative metabolism.

Functionally, BCKDH E2 catalyzes the acyl transfer step of BCAA degradation, converting alpha-ketoacid intermediates into corresponding acyl-CoA products and releasing carbon dioxide. This process provides both energy and metabolic intermediates for biosynthetic pathways. BCKDH E2 activity is tightly regulated by phosphorylation through BCKDH kinase (BCKDK) and dephosphorylation by PPM1K, ensuring proper response to nutrient availability and metabolic stress. In muscle and liver, BCKDH E2 contributes to nitrogen balance, mitochondrial energy generation, and regulation of plasma amino acid levels.

Defects in the DBT gene encoding BCKDH E2 cause Maple Syrup Urine Disease (MSUD) type II, characterized by accumulation of branched-chain amino acids and their ketoacid derivatives, leading to neurological dysfunction and metabolic crisis. Reduced BCKDH E2 function disrupts mitochondrial oxidative decarboxylation, resulting in toxic metabolite buildup. Conversely, increased BCKDH activity enhances amino acid catabolism under fasting or exercise conditions. Pathway associations include branched-chain amino acid degradation, acetyl-CoA biosynthesis, and mitochondrial energy metabolism. BCKDH E2 is highly expressed in liver, skeletal muscle, heart, and brain, reflecting its metabolic importance in energy-demanding tissues.

The BCKDH E2 antibody from NSJ Bioreagents is an ideal reagent for research into amino acid metabolism, mitochondrial enzymology, and inherited metabolic disorders.

Application Notes

Optimal dilution of the BCKDH E2 antibody should be determined by the researcher.

Immunogen

E.coli-derived human DBT recombinant protein (Position: K46-R462) was used as the immunogen for the BCKDH E2 antibody.

Storage

After reconstitution, the BCKDH E2 antibody can be stored for up to one month at 4oC. For long-term, aliquot and store at -20oC. Avoid repeated freezing and thawing.