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BCKDHB Antibody (N-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP10672a

Product Information

Application	WB, IF, FC, IHC-P-Leica, E
Primary Accession	P21953
Other Accession	NP_000047.1
Reactivity	Human, Rat
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB20925
Calculated MW	43122
Antigen Region	41-70

Additional Information

Gene ID	594
Other Names	2-oxoisovalerate dehydrogenase subunit beta, mitochondrial, Branched-chain alpha-keto acid dehydrogenase E1 component beta chain, BCKDE1B, BCKDH E1-beta, BCKDHB
Target/Specificity	This BCKDHB antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 41-70 amino acids from the N-terminal region of human BCKDHB.
Dilution	WB~~1:1000 IF~~1:10~50 FC~~1:10~50 IHC-P-Leica~~1:500 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	BCKDHB Antibody (N-term) is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	BCKDHB (HGNC:987)
Function	Together with BCKDHA forms the heterotetrameric E1 subunit of the

mitochondrial branched-chain alpha-ketoacid dehydrogenase (BCKD) complex. The BCKD complex catalyzes the multi-step oxidative decarboxylation of alpha-ketoacids derived from the branched-chain amino-acids valine, leucine and isoleucine producing CO2 and acyl-CoA which is subsequently utilized to produce energy. The E1 subunit catalyzes the first step with the decarboxylation of the alpha-ketoacid forming an enzyme-product intermediate. A reductive acylation mediated by the lipoylamide cofactor of E2 extracts the acyl group from the E1 active site for the next step of the reaction.

Cellular Location Mitochondrion matrix

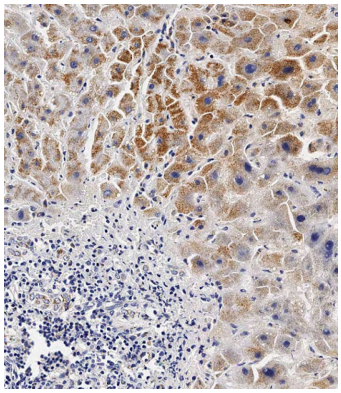
Background

Branched-chain keto acid dehydrogenase is a multienzyme complex associated with the inner membrane of mitochondria, and functions in the catabolism of branched-chain amino acids. The complex consists of multiple copies of 3 components: branched-chain alpha-keto acid decarboxylase (E1), lipoamide acyltransferase (E2) and lipoamide dehydrogenase (E3). This gene encodes the E1 beta subunit, and mutations therein have been associated with maple syrup urine disease (MSUD), type 1B, a disease characterized by a maple syrup odor to the urine in addition to mental and physical retardation, and feeding problems.

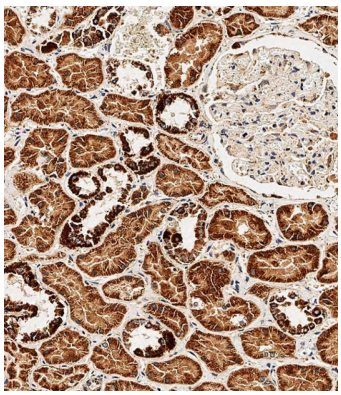
References

Rose, J.E., et al. Mol. Med. 16 (7-8), 247-253 (2010) :
Gorzelany, K., et al. Turk. J. Pediatr. 51(2):97-102(2009)
Quental, S., et al. Mol. Genet. Metab. 94(2):148-156(2008)
Kang, H., et al. Fertil. Steril. 89(3):728-731(2008)
Flaschker, N., et al. J. Inherit. Metab. Dis. 30(6):903-909(2007)

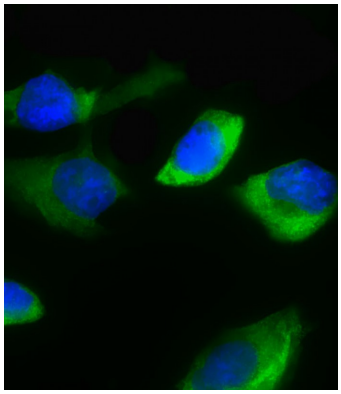
Images



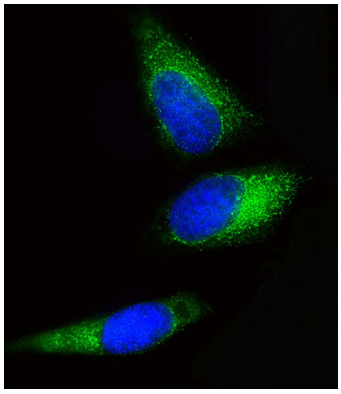
Immunohistochemical analysis of paraffin-embedded Human hepatocarcinoma tissue using AP10672a performed on the Leica® BOND RXm. Tissue was fixed with formaldehyde at room temperature, antigen retrieval was by heat mediation with a EDTA buffer (pH9. 0). Samples were incubated with primary antibody(1:500) for 1 hours at room temperature. A undiluted biotinylated CRF Anti-Polyvalent HRP Polymer antibody was used as the secondary antibody.



Immunohistochemical analysis of paraffin-embedded Human kidney tissue using AP10672a performed on the Leica® BOND RXm. Tissue was fixed with formaldehyde at room temperature, antigen retrieval was by heat mediation with a EDTA buffer (pH9. 0). Samples were incubated with primary antibody(1:500) for 1 hours at room temperature. A undiluted biotinylated CRF Anti-Polyvalent HRP Polymer antibody was used as the secondary antibody.



Immunofluorescent analysis of 4% paraformaldehyde-fixed, 0. 1% Triton X-100 permeabilized Hela cells labeling BCKDHB with AP10672a at 1/25 dilution, followed by Dylight® 488-conjugated goat anti-Rabbit IgG secondary antibody at 1/200 dilution (green). Immunofluorescence image showing Cytoplasm and Weak Nucleus staining on Hela cell line. The nuclear counter stain is DAPI (blue).



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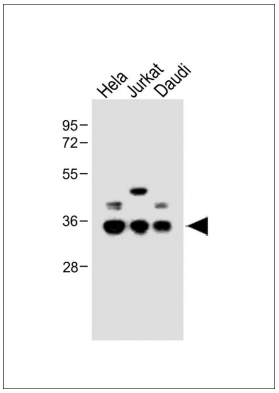
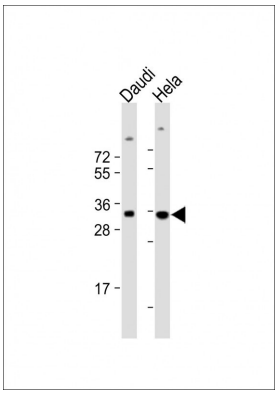


All lanes : Anti-BCKDHB Antibody (N-term) at 1:1000 dilution Lane 1: Hela whole cell lysate Lane 2: Jurkat whole cell lysate Lane 3: Daudi whole cell lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size : 43 kDa Blocking/Dilution buffer: 5% NFDM/TBST.



All lanes : Anti-BCKDHB Antibody (N-term) at 1:1000-1:2000 dilution Lane 1: Daudi whole cell lysate Lane 2: Hela whole cell lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size : 43 kDa Blocking/Dilution buffer: 5% NFDM/TBST.