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ERAB Antibody

Rabbit mAb
Catalog # AP91354

Product Information

Application	WB, IHC, IF, FC, ICC, IHF
Primary Accession	Q99714
Reactivity	Rat, Human, Mouse
Clonality	Monoclonal
Other Names	ABAD; CAMR; HCD2; MHBD; HADH2;
Isotype	Rabbit IgG
Host	Rabbit
Calculated MW	26923

Additional Information

Dilution	WB 1:1000~1:5000 IHC 1:50~1:200 ICC/IF 1:50~1:200 FC 1:50
Purification	Affinity-chromatography
Immunogen	A synthesized peptide derived from human ERAB
Description	Functions in mitochondrial tRNA maturation. Part of mitochondrial ribonuclease P, an enzyme composed of MRPP1/RG9MTD1, MRPP2/HSD17B10 and MRPP3/KIAA0391, which cleaves tRNA molecules in their 5'-ends. By interacting with intracellular amyloid-beta, it may contribute to the neuronal dysfunction associated with Alzheimer disease (AD).
Storage Condition and Buffer	Rabbit IgG in phosphate buffered saline , pH 7.4, 150mM NaCl, 0.02% sodium azide and 50% glycerol. Store at +4°C short term. Store at -20°C long term. Avoid freeze / thaw cycle.

Protein Information

Name	HSD17B10
Synonyms	ERAB, HADH2, MRPP2, SCHAD, SDR5C1, XH98G
Function	Mitochondrial dehydrogenase involved in pathways of fatty acid, branched-chain amino acid and steroid metabolism (PubMed:10600649, PubMed:12917011, PubMed:18996107, PubMed:19706438, PubMed:20077426, PubMed:25925575, PubMed:26950678, PubMed:28888424, PubMed:9553139). Acts as (S)-3-hydroxyacyl-CoA dehydrogenase in mitochondrial fatty acid beta-oxidation, a major degradation pathway of fatty acids. Catalyzes the third step in the beta-oxidation cycle, namely the reversible conversion of (S)-3-hydroxyacyl-CoA to 3- ketoacyl-CoA. Preferentially accepts straight medium- and short-chain acyl-CoA substrates with highest efficiency for (3S)-hydroxybutanoyl- CoA (PubMed:10600649, PubMed:12917011, PubMed:25925575, PubMed:26950678, PubMed:9553139). Acts as

3-hydroxy-2-methylbutyryl-CoA dehydrogenase in branched-chain amino acid catabolic pathway. Catalyzes the oxidation of 3-hydroxy-2-methylbutanoyl-CoA into 2-methyl-3- oxobutanoyl-CoA, a step in isoleucine degradation pathway (PubMed:18996107, PubMed:19706438, PubMed:20077426). Has hydroxysteroid dehydrogenase activity toward steroid hormones and bile acids. Catalyzes the oxidation of 3alpha-, 17beta-, 20beta- and 21- hydroxysteroids and 7alpha- and 7beta-hydroxy bile acids (PubMed:10600649, PubMed:12917011). Oxidizes allopregnanolone/brexanolone at the 3alpha-hydroxyl group, which is known to be critical for the activation of gamma-aminobutyric acid receptors (GABAARs) chloride channel (PubMed:19706438, PubMed:28888424). Has phospholipase C-like activity toward cardiolipin and its oxidized species. Likely oxidizes the 2'-hydroxyl in the head group of cardiolipin to form a ketone intermediate that undergoes nucleophilic attack by water and fragments into diacylglycerol, dihydroxyacetone and orthophosphate. Has higher affinity for cardiolipin with oxidized fatty acids and may degrade these species during the oxidative stress response to protect cells from apoptosis (PubMed:26338420). By interacting with intracellular amyloid-beta, it may contribute to the neuronal dysfunction associated with Alzheimer disease (AD) (PubMed:9338779). Essential for structural and functional integrity of mitochondria (PubMed:20077426).

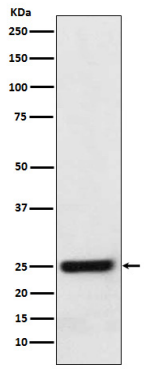
Cellular Location

Mitochondrion. Mitochondrion matrix, mitochondrion nucleoid

Tissue Location

Ubiquitously expressed in normal tissues but is overexpressed in neurons affected in AD.

Images



Western blot analysis of ERAB expression in HeLa cell lysate.

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