SANTA CRUZ BIOTECHNOLOGY, INC.

ETFDH (C-18): sc-242642

BACKGROUND

ETFDH (electron-transferring-flavoprotein dehydrogenase), also known as electron transfer flavoprotein-ubiquinone oxidoreductase, MADD or ETFQO, is a 617 amino acid membrane-bound electron transfer protein that exists as a monomer, localizes to the mitochondrial inner membrane and belongs to the ETF-QO/fixC family. ETFDH accepts electrons from electron-transfer flavoprotein (ETF) in the mitochondrial matrix while reducing ubiquinone in the mitochondrial membrane. ETFDH is encoded by a gene mapping to human chromosome 4q32.1, and contains one molecule of FAD and a 4Fe-4S cluster. As a result of alternative splicing events, two ETFDH isoforms exist. Defects in ETFDH are responsible for an autosomal recessive disorder of amino acid, fatty acid and choline metabolism, known as glutaric aciduria type 2C (GA2C) or multiple acyl-CoA dehydrogenation deficiency (MADD). GA2C is characterized by severe hypoketotic hypoglycemia and acidosis.

REFERENCES

1. Lehnert, W., et al. 1982. Multiple acyl-CoA dehydrogenation deficiency (glutaric aciduria type II), congenital polycystic kidneys, and symmetric warty dysplasia of the cerebral cortex in two brothers. I. Clinical, metabolical, and biochemical findings. Eur. J. Pediatr. 139: 56-59.
2. Böhm, N., et al. 1982. Multiple acyl-CoA dehydrogenation deficiency (glutaric aciduria type II), congenital polycystic kidneys, and symmetric warty dysplasia of the cerebral cortex in two newborn brothers. II. Morphology and pathogenesis. Eur. J. Pediatr. 139: 60-65.
3. White, R.A., et al. 1996. Assignment of Etfdh, Etfb, and Etfa to chromosomes 3, 7, and 13: the mouse homologs of genes responsible for glutaric acidemia type II in human. Genomics 33: 131-134.
4. Olsen, R.K., et al. 2003. Clear relationship between ETF/ETFDH genotype and phenotype in patients with multiple acyl-CoA dehydrogenation deficiency. Hum. Mutat. 22: 12-23.
5. Online Mendelian Inheritance in Man, OMIM™. 2005. Johns Hopkins University, Baltimore, MD. MIM Number: 231675. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/
6. Olsen, R.K., et al. 2007. ETFDH mutations as a major cause of riboflavin-responsive multiple acyl-CoA dehydrogenation deficiency. Brain 130 (Pt. 8): 2045-2054.

CHROMOSOMAL LOCATION

Genetic locus: ETFDH (human) mapping to 4q32.1; Etfdh (mouse) mapping to 3 E3.

SOURCE

ETFDH (C-18) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of ETFDH of human origin.

STORAGE

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

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-242642 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

ETFDH (C-18) is recommended for detection of ETFDH of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg 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).

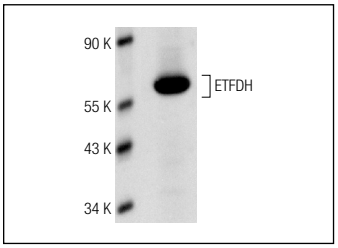
ETFDH (C-18) is also recommended for detection of ETFDH in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for ETFDH siRNA (h): sc-89048, ETFDH siRNA (m): sc-144955, ETFDH shRNA Plasmid (h): sc-89048-SH, ETFDH shRNA Plasmid (m): sc-144955-SH, ETFDH shRNA (h) Lentiviral Particles: sc-89048-V and ETFDH shRNA (m) Lentiviral Particles: sc-144955-V.

Molecular Weight of ETFDH: 68 kDa.

Positive Controls: mouse heart extract: sc-2254.

DATA



ETFDH (C-18): sc-242642. Western blot analysis of ETFDH expression in mouse heart tissue extract.

RESEARCH USE

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

PROTOCOLS

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