

# ETFDH (D-2): sc-515202

## 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, metabolic, 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.

## CHROMOSOMAL LOCATION

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

## SOURCE

ETFDH (D-2) is a mouse monoclonal antibody raised against amino acids 302-600 mapping near the C-terminus of ETFDH of human origin.

## PRODUCT

Each vial contains 200 µg IgG<sub>1</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

ETFDH (D-2) is available conjugated to agarose (sc-515202 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-515202 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-515202 PE), fluorescein (sc-515202 FITC), Alexa Fluor® 488 (sc-515202 AF488), Alexa Fluor® 546 (sc-515202 AF546), Alexa Fluor® 594 (sc-515202 AF594) or Alexa Fluor® 647 (sc-515202 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-515202 AF680) or Alexa Fluor® 790 (sc-515202 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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## STORAGE

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

## APPLICATIONS

ETFDH (D-2) is recommended for detection of ETFDH of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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).

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.

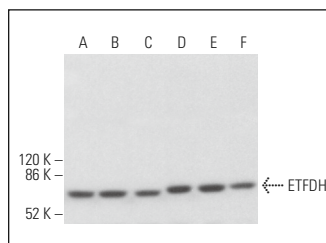
Positive Controls: Jurkat whole cell lysate: sc-2204, HuT 78 whole cell lysate: sc-2208 or Ramos cell lysate: sc-2216.

## RECOMMENDED SUPPORT REAGENTS

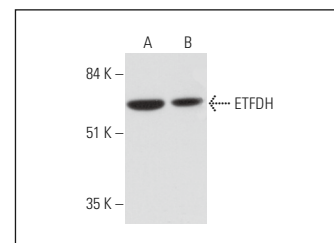
To ensure optimal results, the following support reagents are recommended:

- 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048.
- 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).
- 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

## DATA



ETFDH (D-2): sc-515202. Western blot analysis of ETFDH expression in Jurkat (A), Raji (B), BJAB (C), BYDP (D), RAW 264.7 (E) and 3611-RF (F) whole cell lysates.



ETFDH (D-2): sc-515202. Western blot analysis of ETFDH expression in HuT 78 (A) and Ramos (B) whole cell lysates.

## SELECT PRODUCT CITATIONS

1. Tiwari, S., et al. 2020. Gender-specific changes in energy metabolism and protein degradation as major pathways affected in livers of mice treated with ibuprofen. *Sci. Rep.* 10: 3386.
2. Xiao, C., et al. 2020. Mitochondrial energetic impairment in a patient with late-onset glutaric acidemia type 2. *Am. J. Med. Genet. A* 182: 2426-2431.

## RESEARCH USE

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