

fumarate hydratase (H-6): sc-393992

BACKGROUND

Fumarate hydratase, a ubiquitously expressed mitochondrial enzyme, catalyses the reversible hydration of fumaric acid to yield L-malic acid during the Krebs cycle. Germline mutations in the fumarate hydratase gene cause a predisposition to renal defects such as multiple cutaneous and uterine leiomyoma (MCL) syndrome. Furthermore, mutations also correlate with renal and smooth muscle tumors, but not with prostate cancer. Additionally, like other metabolic diseases, fumarate hydratase deficiency correlates with seizures, due to prenatal brain dysgenesis.

CHROMOSOMAL LOCATION

Genetic locus: FH (human) mapping to 1q43; Fh1 (mouse) mapping to 1 H4.

SOURCE

fumarate hydratase (H-6) is a mouse monoclonal antibody raised against amino acids 426-510 mapping at the C-terminus of fumarate hydratase of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

fumarate hydratase (H-6) is available conjugated to agarose (sc-393992 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-393992 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-393992 PE), fluorescein (sc-393992 FITC), Alexa Fluor® 488 (sc-393992 AF488), Alexa Fluor® 546 (sc-393992 AF546), Alexa Fluor® 594 (sc-393992 AF594) or Alexa Fluor® 647 (sc-393992 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-393992 AF680) or Alexa Fluor® 790 (sc-393992 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

RESEARCH USE

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

APPLICATIONS

fumarate hydratase (H-6) is recommended for detection of fumarate hydratase 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), immunohistochemistry (including paraffin-embedded sections) (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 fumarate hydratase siRNA (h): sc-105377, fumarate hydratase siRNA (m): sc-145272, fumarate hydratase shRNA Plasmid (h): sc-105377-SH, fumarate hydratase shRNA Plasmid (m): sc-145272-SH, fumarate hydratase shRNA (h) Lentiviral Particles: sc-105377-V and fumarate hydratase shRNA (m) Lentiviral Particles: sc-145272-V.

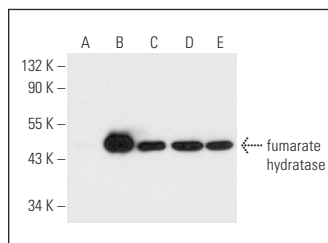
Molecular Weight of fumarate hydratase: 46 kDa.

Positive Controls: fumarate hydratase (m): 293T Lysate: sc-120338, HeLa whole cell lysate: sc-2200 or Hep G2 cell lysate: sc-2227.

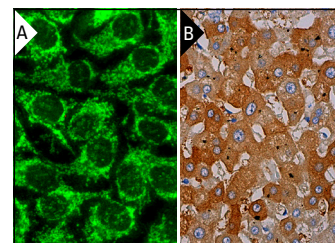
RECOMMENDED SUPPORT REAGENTS

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. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



fumarate hydratase (H-6): sc-393992. Western blot analysis of fumarate hydratase expression in non-transfected 293T: sc-117752 (A), mouse fumarate hydratase transfected 293T: sc-120338 (B), HeLa (C), Hep G2 (D) and A-431 (E) whole cell lysates.



fumarate hydratase (H-6): sc-393992. Immunofluorescence staining of methanol-fixed HeLa cells showing mitochondrial localization (A). Immunoperoxidase staining of formalin fixed, paraffin-embedded human liver tissue showing cytoplasmic staining of hepatocytes (B).

SELECT PRODUCT CITATIONS

- Gibson, G.E., et al. 2015. α-ketoglutarate dehydrogenase complex-dependent succinylation of proteins in neurons and neuronal cell lines. *J. Neurochem.* 134: 86-96.
- Drusian, L., et al. 2018. MTORC1 upregulation leads to accumulation of the oncometabolite fumarate in a mouse model of renal cell carcinoma. *Cell Rep.* 24: 1093-1104.e6.
- Xu, Y., et al. 2018. Pathologic oxidation of PTPN12 underlies ABL1 phosphorylation in hereditary leiomyomatosis and renal cell carcinoma. *Cancer Res.* 78: 6539-6548.
- Urizar-Arenaza, I., et al. 2020. SPANX-A/D protein subfamily plays a key role in nuclear organisation, metabolism and flagellar motility of human spermatozoa. *Sci. Rep.* 10: 5625.
- Cai, Z., et al. 2020. Phosphorylation of PDHA by AMPK drives TCA cycle to promote cancer metastasis. *Mol. Cell* 80: 263-278.e7.

STORAGE

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

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