SANTA CRUZ BIOTECHNOLOGY, INC.

fumarate hydratase (J-13): sc-100743

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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.

SOURCE

fumarate hydratase (J-13) is a mouse monoclonal antibody raised against recombinant fumarate hydratase of human origin.

PRODUCT

Each vial contains 100 µg IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

fumarate hydratase (J-13) is recommended for detection of precursor mitochondrial and cytoplasmic mature chains of fumarate hydratase of 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), 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 shRNA Plasmid (h): sc-105377-SH and fumarate hydratase shRNA (h) Lentiviral Particles: sc-105377-V.

Molecular Weight of fumarate hydratase: 46 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200.

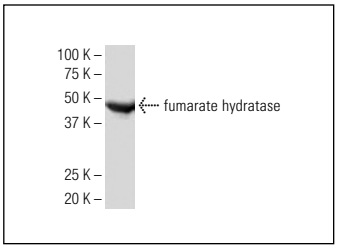
RECOMMENDED SUPPORT REAGENTS

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.

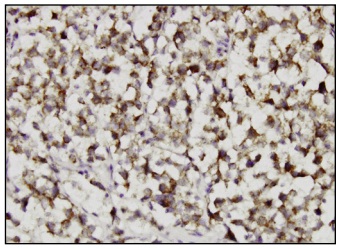
STORAGE

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

DATA



fumarate hydratase (J-13): sc-100743. Western blot analysis of fumarate hydratase expression in HeLa whole cell lysate.



fumarate hydratase (J-13): sc-100743. Immunoperoxidase staining of formalin-fixed, paraffin-embedded human seminoma tissue showing cytoplasmic localization.

SELECT PRODUCT CITATIONS

1. Sartor, F., et al. 2012. Adaptive metabolic response to 4 weeks of sugar-sweetened beverage consumption in healthy, lightly active individuals and chronic high glucose availability in primary human myotubes. Eur. J. Nutr. 52: 937-948.
2. Martínek, P., et al. 2015. Genetic testing of leiomyoma tissue in women younger than 30 years old might provide an effective screening approach for the hereditary leiomyomatosis and renal cell cancer syndrome (HLRCC). Virchows Arch. 467: 185-191.
3. Harrison, W.J., et al. 2016. fumarate hydratase-deficient uterine leiomyomas occur in both the syndromic and sporadic settings. Am. J. Surg. Pathol. 40: 599-607.
4. Bell, R.C., et al. 2016. Rare leiomyoma of the tunica dartos: a case report with clinical relevance for malignant transformation and HLRCC. Case Rep. Pathol. 2016: 6471520.
5. Trpkov, K., et al. 2016. fumarate hydratase-deficient renal cell carcinoma is strongly correlated with fumarate hydratase mutation and hereditary leiomyomatosis and renal cell carcinoma syndrome. Am. J. Surg. Pathol. 40: 865-875.
6. Bortoletto, P., et al. 2017. Hereditary leiomyomatosis and renal cell cancer: cutaneous lesions & atypical fibroids. Case Rep. Womens Health 15: 31-34.
7. Li, Y., et al. 2018. Re-evaluation of 33 "unclassified" eosinophilic renal cell carcinomas in young patients. Histopathology 72: 588-600.
8. Muller, M., et al. 2018. Pattern multiplicity and fumarate hydratase (FH)/S-(2-succino)-cysteine (2SC) staining but not eosinophilic nucleoli with perinucleolar halos differentiate hereditary leiomyomatosis and renal cell carcinoma-associated renal cell carcinomas from kidney tumors without FH gene alteration. Mod. Pathol. 31: 974-983.

RESEARCH USE

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