



# SZABO SCANDIC

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## Produktinformation



Forschungsprodukte & Biochemikalien



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Diagnostik & molekulare Diagnostik



Laborgeräte & Service

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### Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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# fumarate hydratase (m): 293T Lysate: sc-120338

## BACKGROUND

Fumarate hydratase, a ubiquitously expressed mitochondrial enzyme, catalyzes 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.

## REFERENCES

1. De Meirleir, L. 2002. Defects of pyruvate metabolism and the Krebs cycle. *J. Child Neurol. Suppl. 3:* 3S26-3S34.
2. Eng, C., et al. 2003. A role for mitochondrial enzymes in inherited neoplasia and beyond. *Nat. Rev. Cancer 3:* 193-202.
3. Martinez-Mir, A., et al. 2003. Germline fumarate hydratase mutations in families with multiple cutaneous and uterine leiomyomata. *J. Invest. Dermatol. 121:* 741-744.
4. Bevan, S., et al. 2003. Germline mutations in fumarate hydratase (FH) do not predispose to prostate cancer. *Prostate Cancer Prostatic Dis. 6:* 12-14.
5. Pollard, P.J., et al. 2003. The TCA cycle and tumorigenesis: the examples of fumarate hydratase and succinate dehydrogenase. *Ann. Med. 35:* 632-639.
6. Gross, K.L., et al. 2004. Involvement of fumarate hydratase in nonsyndromic uterine leiomyomas: genetic linkage analysis and fish studies. *Genes Chromosomes Cancer 41:* 183-190.
7. McGrath, J.A. 2003. Biologic lessons from mutations in the Krebs cycle enzyme, fumarate hydratase. *J. Invest. Dermatol. 121:* vii.

## CHROMOSOMAL LOCATION

Genetic locus: Fh1 (mouse) mapping to 1 H4.

## PRODUCT

fumarate hydratase (m): 293T Lysate represents a lysate of mouse fumarate hydratase transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

## APPLICATIONS

fumarate hydratase (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive fumarate hydratase antibodies. Recommended use: 10-20 µl per lane.

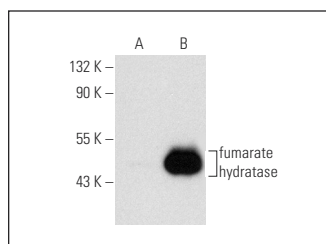
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

fumarate hydratase (H-6): sc-393992 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse fumarate hydratase expression in fumarate hydratase transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

## 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.

## DATA



fumarate hydratase (H-6): sc-393992. Western blot analysis of fumarate hydratase expression in non-transfected: sc-117752 (A) and mouse fumarate hydratase transfected: sc-120338 (B) 293T whole cell lysates.

## STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

## RESEARCH USE

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

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) for detailed protocols and support products.