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.

REFERENCES

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 IgG2b in 1.0 ml PBS with < 0.1% sodium azide and 0.1% gelatin.

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

RECOMMENDED SECONDARY REAGENTS
To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-mouse IgG-HRP: sc-2005 (dilution range: 1:2000-1:32,000) or Cruz Marker™ compatible goat anti-mouse IgG-HRP: sc-2031 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 goat anti-mouse IgG-FITC: sc-2010 (dilution range: 1:100-1:400) or goat anti-mouse IgG-TR: sc-2781 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941. 4) Immunohistochemistry: use ImmunoCruz™: sc-2050 or ABC: sc-2017 mouse IgG Staining Systems.

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).

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