

# Human Fumarase/FH antibody

Catalog Number: ATGA0366

## PRODUCT INFORMATION

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**Catalog number**

ATGA0366

**Clone No.**

AT3F6

**Product type**

Monoclonal Antibody

**UnitProt No.**

P07954

**NCBI Accession No.**

NP\_000134

**Alternative Names**

Fumarate hydratase, FH, HLRCC, LRCC, MCL, MCuL1, Fumarate hydratase Fumarase, Fumarate hydratase mitochondrial, MCuL 1, Multiple hereditary cutaneous leiomyomata.

## PRODUCT SPECIFICATION

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**Antibody Host**

Mouse

**Reacts With**

Human

**Concentration**

1mg/ml (determined by BCA assay)

**Formulation**

Liquid in. Phosphate-Buffered Saline (pH 7.4) with 0.02% Sodium Azide, 10% glycerol

**Immunogen**

Recombinant human Fumarase (44-510aa) purified from E. coli

**Isotype**

IgG2a kappa

**Purification Note**

By protein-A affinity chromatography

**Application**

ELISA, WB, ICC/IF, FACS

**Usage**

The antibody has been tested by ELISA and Western blot analysis to assure specificity and reactivity. Since application varies, however, each investigation should be titrated by the reagent to obtain optimal results.

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## Storage

Can be stored at +2C to +8C for 1 week. For long term storage, aliquot and store at -20C to -80C. Avoid repeated freezing and thawing cycles.

## BACKGROUND

### Description

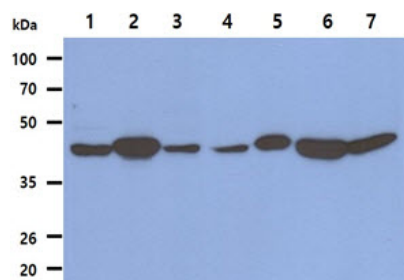
Fumarase (Fumarate hydratase) is an enzyme that catalyzes the reversible hydration/dehydration of fumarate to S-malate and is involved in the tricarboxylic acid (TCA), or Krebs cycle. This enzyme exists in both a cytosolic form and an N-terminal extended mitochondrial form. The N-terminal extended form is targeted to the mitochondrion, where the removal of the extension is the same form as in the cytoplasm. Fumarase deficiency can lead to progressive encephalopathy, cerebral atrophy and developmental delay and this enzyme also is thought to act as a tumor suppressor.

### General References

Lehtonen R., et al. (2003) J Med Genet. 40(3): e19.  
Toro JR., et al. (2003) Am J Hum Genet. 73(1): 95-106.

## DATA

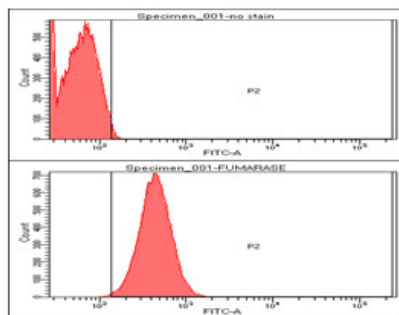
### Western blot analysis (WB)



The Cell lysates (40ug) were resolved by SDS-PAGE, transferred to PVDF membrane and probed with anti-human Fumarase antibody (1:1000). Proteins were visualized using a goat anti-mouse secondary antibody conjugated to HRP and an ECL detection system.

Lane 1. : HeLa cell lysate  
Lane 2. : LNCap cell lysate  
Lane 3. : A549 cell lysate  
Lane 4. : HepG2 cell lysate  
Lane 5. : 293T cell lysate  
Lane 6. : K562 cell lysate  
Lane 7. : Jurkat cell lysate

### Flow cytometry (FACS)



Flow cytometry analysis of Fumarase in HeLa cell line, staining at 2-5ug for 1x10<sup>6</sup> cells. The secondary antibody used goat anti-mouse IgG Alexa fluor 488 conjugate.