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# FH Antibody (N-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP18400a

## Product Information

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| **Application** | WB, E |
| **Primary Accession** | P07954 |
| **Other Accession** | P14408, P10173, P97807, Q60HF9, NP_000134.2 |
| **Reactivity** | Human, Rat, Mouse |
| **Predicted** | Monkey, Mouse, Pig, Rat |
| **Host** | Rabbit |
| **Clonality** | Polyclonal |
| **Isotype** | Rabbit IgG |
| **Clone Names** | RB38443 |
| **Calculated MW** | 54637 |
| **Antigen Region** | 107-135 |

## Additional Information

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| **Gene ID** | 2271 |
| **Other Names** | Fumarate hydratase, mitochondrial, Fumarase, FH |
| **Target/Specificity** | This FH antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 107-135 amino acids from the N-terminal region of human FH. |
| **Dilution** | WB~~1:1000 E~~Use at an assay dependent concentration. |
| **Format** | Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification. |
| **Storage** | Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles. |
| **Precautions** | FH Antibody (N-term) is for research use only and not for use in diagnostic or therapeutic procedures. |

## Protein Information

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| **Name** | FH {ECO:0000303\|PubMed:27037871, ECO:0000312\|HGNC:HGNC:3700} |
| **Function** | Catalyzes the reversible stereospecific interconversion of fumarate to L-malate (PubMed:30761759). Experiments in other species have demonstrated that specific isoforms of this protein act in defined pathways |

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| | and favor one direction over the other (Probable). |
| **Cellular Location** | [Isoform Mitochondrial]: Mitochondrion |
| **Tissue Location** | Expressed in red blood cells; underexpressed in red blood cells (cytoplasm) of patients with hereditary non-spherocytic hemolytic anemia of unknown etiology. |

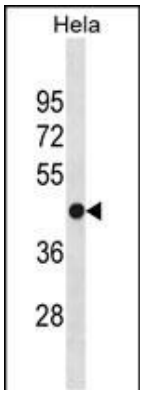
## Background

The protein encoded by this gene is an enzymatic component of the tricarboxylic acid (TCA) cycle, or Krebs cycle, and catalyzes the formation of L-malate from fumarate. It exists in both a cytosolic form and an N-terminal extended form, differing only in the translation start site used. The N-terminal extended form is targeted to the mitochondrion, where the removal of the extension generates the same form as in the cytoplasm. It is similar to some thermostable class II fumarases and functions as a homotetramer. Mutations in this gene can cause fumarase deficiency and lead to progressive encephalopathy.

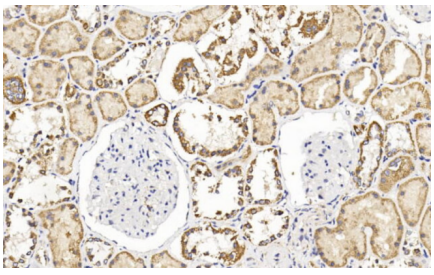
## References


Shimada, M., et al. Hum. Genet. 128(4):433-441(2010)
Allegri, G., et al. J. Inherit. Metab. Dis. 33(4):411-419(2010)
Yogev, O., et al. PLoS Biol. 8 (3), E1000328 (2010) :
Yang, Y., et al. Cancer Genet. Cytogenet. 196(1):45-55(2010)
Rikova, K., et al. Cell 131(6):1190-1203(2007)


## Images




FH Antibody (N-term) (Cat. #AP18400a) western blot analysis in Hela cell line lysates (35ug/lane).This demonstrates the FH Antibody detected the FH protein (arrow).



Immunohistochemical analysis of paraffin-embedded Human kidney section using Pink1(Cat#AP18400a). AP18400a was diluted at 1:800 dilution. A undiluted biotinylated goat polyvalent antibody was used as the secondary, followed by DAB staining.