

AP21256AF-N**Polyclonal Antibody to Fumarase - Azide Free**

Alternate names:	HLRCC, LRCC, MCL, MCUL1, mitochondrial Fumarate hydratase
Quantity:	10 mg
Concentration:	10.0 mg/ml
Background:	Defects in FH are the cause of fumarase deficiency (FHD) [MIM:606812]; also known as fumaricaciduria. FHD is characterized by progressive encephalopathy, developmental delay, hypotonia, cerebral atrophy and lactic and pyruvic acidemia. Defects in FH are the cause of multiple cutaneous and uterine leiomyomata (MCUL1) [MIM:150800]. MCUL1 is an autosomal dominant condition in which affected individuals develop benign smooth muscle tumors (leiomyomata) of the skin. Affected females also usually develop leiomyomata of the uterus (fibroids). Defects in FH are the cause of hereditary leiomyomatosis and renal cell cancer (HLRCC) [MIM:605839].
Uniprot ID:	P10173
NCBI:	9823
Host / Isotype:	Rabbit / IgG
Immunogen:	Fumarase is isolated and purified from Porcine heart Freund's complete adjuvant is used in the first step of the immunization procedure.
Format:	State: Lyophilized hyperimmune IgG fraction Purification: Ammonium Sulphate Precipitation and Ion Exchange Chromatography Buffer System: PBS, pH 7.2 without preservatives and foreign proteins Reconstitution: Restore by adding 1.0 ml of sterile distilled water
Applications:	This product is intended for use in precipitating and non-precipitating antibody-binding assays (such as e.g., ELISA and Western blotting and Immunofluorescence or Histochemical techniques), to prepare an insoluble immuno-affinity adsorbent, for labelling with a marker of the customer's own choice. Working dilutions in non-precipitating antibody-binding techniques: 1/1,000-1/5,000. Other applications not tested. Optimal dilutions are dependent on conditions and should be determined by the user.
Specificity:	The antibody recognizes Fumarase from Porcine heart. The reagents were evaluated for potency, purity and specificity using most or all of the following techniques: Immunoelectrophoresis, Cross-Immunoelectrophoresis, single Radial Immunodiffusion (Ouchterlony), block titration, ELISA, Immunoblotting and Enzyme Inhibition. Cross-reactivities against enzymes of other sources may occur but have not been determined. Species: Pig. Other species not tested.

Storage:

Store the antibody lyophilized at 2-8°C and reconstituted at 2-8°C for one week or (in aliquots) at -20°C for longer.

If a slight precipitation occurs upon storage, this should be removed by centrifugation.

Shelf life: one year from despatch.