

**Mouse Anti-Fumarate Hydratase (FH) [J13]: MC0434, MC0434RTU7**

**Intended Use:** For Research Use Only

**Description:** Fumarate hydratase (FH) or fumarase encoded by the FH gene and 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. FH and succinate dehydrogenase are tumour suppressors and they are associated with metabolic dysfunction and tumorigenesis, providing biochemical evidence to explain enhanced glycolysis in tumours. Hereditary Leiomyomatosis and Renal Cell Cancer (HLRCC) is an autosomal dominant heritable syndrome with predisposition to development of Renal Cell Carcinoma and Smooth Muscle Tumors of the skin and uterus. Cells of individuals with HLRCC had lower fumarate hydratase antibody activity than cells from normal controls, making fumarate hydratase antibody activity testing a useful method for diagnosis and screening.

**Specifications**

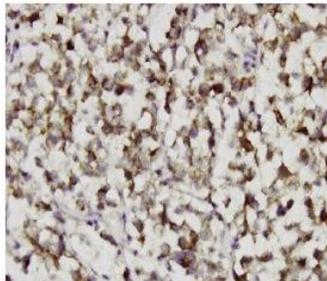
Clone: J13  
 Source: Mouse  
 Isotype: IgG2b/k  
 Reactivity: Human  
 Immunogen: Recombinant fumarate hydratase of human origin  
 Localization: Cytoplasm  
 Formulation: Antibody in PBS pH7.4, containing BSA and  $\leq 0.09\%$  sodium azide (NaN<sub>3</sub>)  
 Storage: Store at 2°- 8°C  
 Applications: IHC, ELISA, IF, IP, WB  
 Package:

| Description                          | Catalog No. | Size |
|--------------------------------------|-------------|------|
| Fumarate Hydratase (FH) Concentrated | MC0434      | 1 ml |
| Fumarate Hydratase (FH) Prediluted   | MC0434RTU7  | 7 ml |

**IHC Procedure\***

Positive Control Tissue: Kidney and clear cell renal cancer; HeLa and NIH/3T3 cells  
 Concentrated Dilution: 50-200  
 Pretreatment: Tris EDTA pH9.0, 15 minutes using Pressure Cooker, or 30-60 minutes using water bath at 95°-99°C  
 Incubation Time and Temp: 30-60 minutes @ RT  
 Detection: Refer to the detection system manual

\* Result should be confirmed by an established diagnostic procedure.



FFPE human seminoma tissue stained with anti-FH using DAB

**References:**

1. Comprehensive Molecular Characterization and Response to Therapy in Fumarate Hydratase-Deficient Renal Cell Carcinoma. Gleeson, JP. et al. Clin Cancer Res. 2021.