

## Mouse Anti-Fumarase/FH antibody

SLM-33167M

**Product Name** Fumarase/FH

**Chinese Name** 富马酸水合酶单克隆抗体

**Alias** FH; Fumarase; Fumarate hydratase; Fumarate hydratase mitochondrial; FUMH\_HUMAN; HLRCC; LRCC; MCL; MCUL 1; MCUL1; Multiple hereditary cutaneous leiomyomata.

**Research Area** Tumour Cell biology immunology transcriptional regulatory factor Mitochondrion

**Immunogen Species** Mouse

**Clonality** Monoclonal

**Clone NO.** 4C12

**React Species** Human,Mouse,Rat

**Applications** IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,WB=1:500-3000 (Paraffin sections need antigen repair)  
not yet tested in other applications.  
optimal dilutions/concentrations should be determined by the end user.

**Theoretical molecular weight** 51kDa

**Cellular localization** cytoplasmic Mitochondrion

**Form** Liquid

**Concentration** 1mg/ml

**immunogen** Recombinant human Fumarase/FH Protein

**Lsotype** IgG

**Purification** affinity purified by Protein G

**Buffer Solution** Human,Mouse,Rat1M TBS(pH7.4) with 1% BSA, Human,Mouse,Rat3% Proclin300 and 50% Glycerol.

**Storage** Shipped at 4°C. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.

**Attention** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

**PubMed** [PubMed](#)

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. [provided by RefSeq, Jul 2008]

**Function:**

Also acts as a tumor suppressor.

**Subunit:**

Homotetramer.

**Subcellular Location:**

Cytoplasm, Mitochondrion

**Tissue Specificity:**

Expressed in red blood cells; underexpressed in red blood cells (cytoplasm) of patients with hereditary non-spherocytic hemolytic anemia of unknown etiology.

**Product  
Detail**

**DISEASE:**

Fumarase deficiency (FMRD): The disease is caused by mutations affecting the gene represented in this entry. A severe autosomal recessive metabolic disorder characterized by early-onset hypotonia, profound psychomotor retardation, and brain abnormalities, such as agenesis of the corpus callosum, gyral defects, and ventriculomegaly. Many patients show neonatal distress, metabolic acidosis, and/or encephalopathy.

Hereditary leiomyomatosis and renal cell cancer (HLRCC): The disease is caused by mutations affecting the gene represented in this entry. A disorder characterized by predisposition to cutaneous and uterine leiomyomas, and papillary type 2 renal cancer which occurs in about 20% of patients.

**Similarity:**

Belongs to the class-II fumarase/aspartase family. Fumarase subfamily.

**SWISS:**

P07954

**Gene ID:**

2271

**Database links:**

[Entrez Gene: 520260](#) Cow

[Entrez Gene: 480092](#) Dog

[Entrez Gene: 2271](#) Human

[Entrez Gene: 14194](#) Mouse

[Entrez Gene: 100627128](#) Pig

[Entrez Gene: 24368](#) Rat

[Entrez Gene: 393938](#) Zebrafish

[Omim: 136850](#) Human

[SwissProt: P07954](#) Human

[SwissProt: P97807](#) Mouse

[SwissProt: P10173](#) Pig

[SwissProt: P14408](#) Rat

[SwissProt: Q7SX99](#) Zebrafish

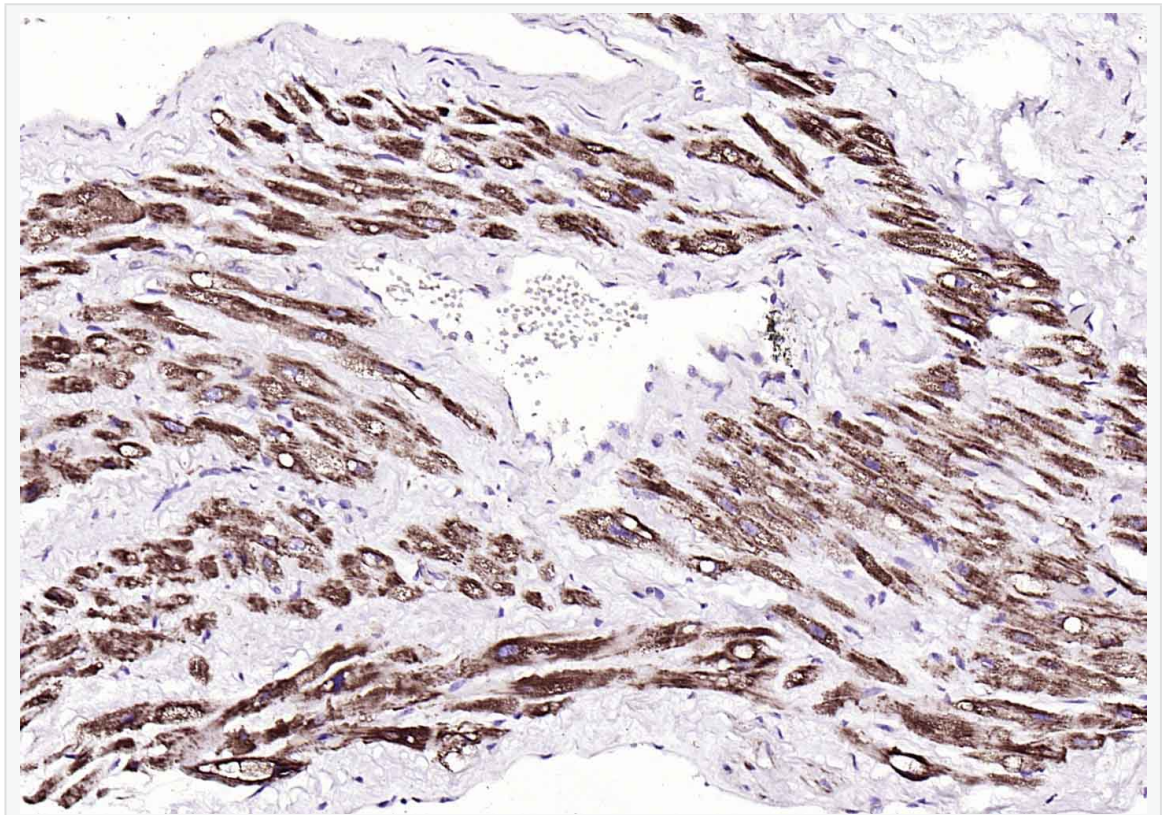
[Unigene: 592490](#) Human

[Unigene: 41502](#) Mouse

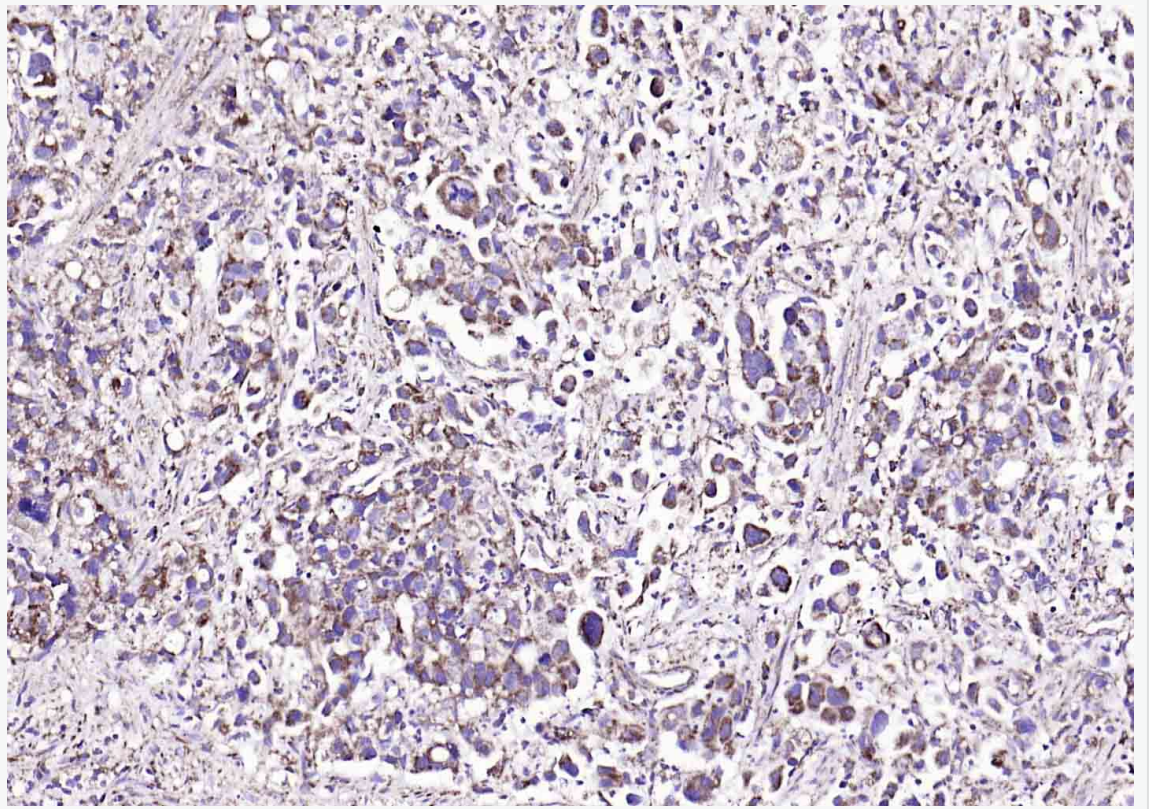
[Unigene: 29782](#) Rat

[Unigene: 104452](#) Zebrafish

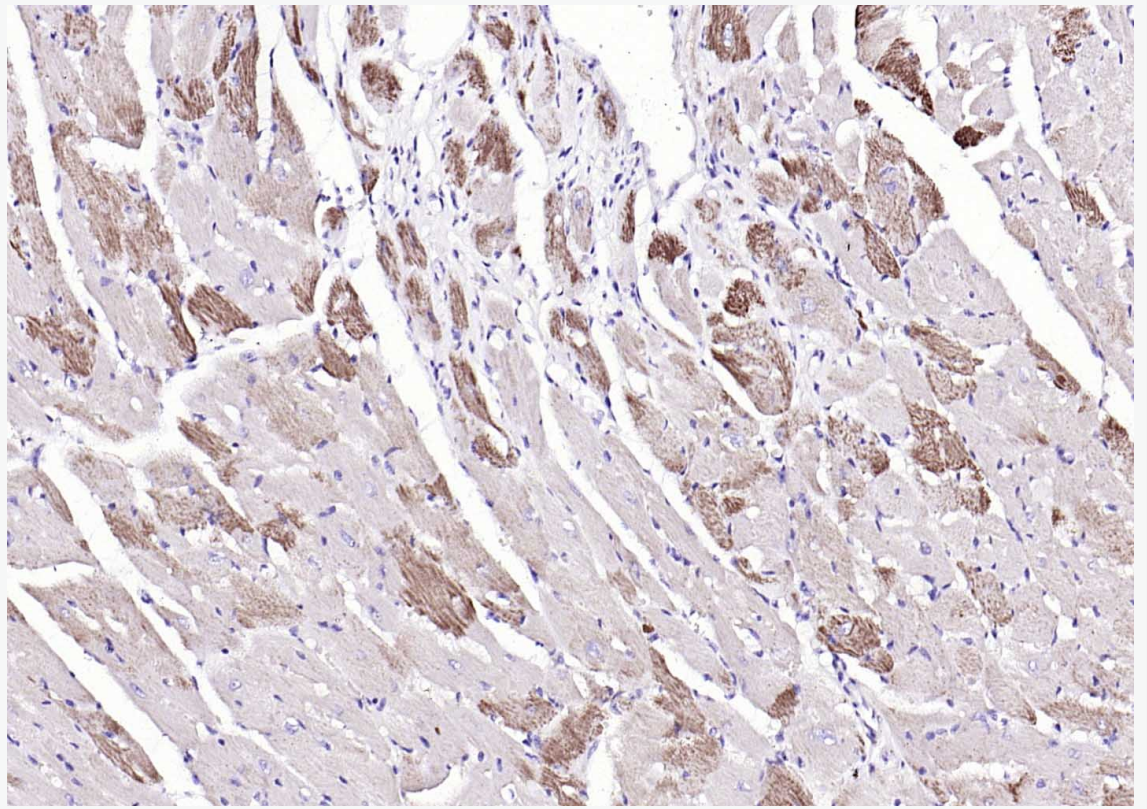
**Product  
Picture**



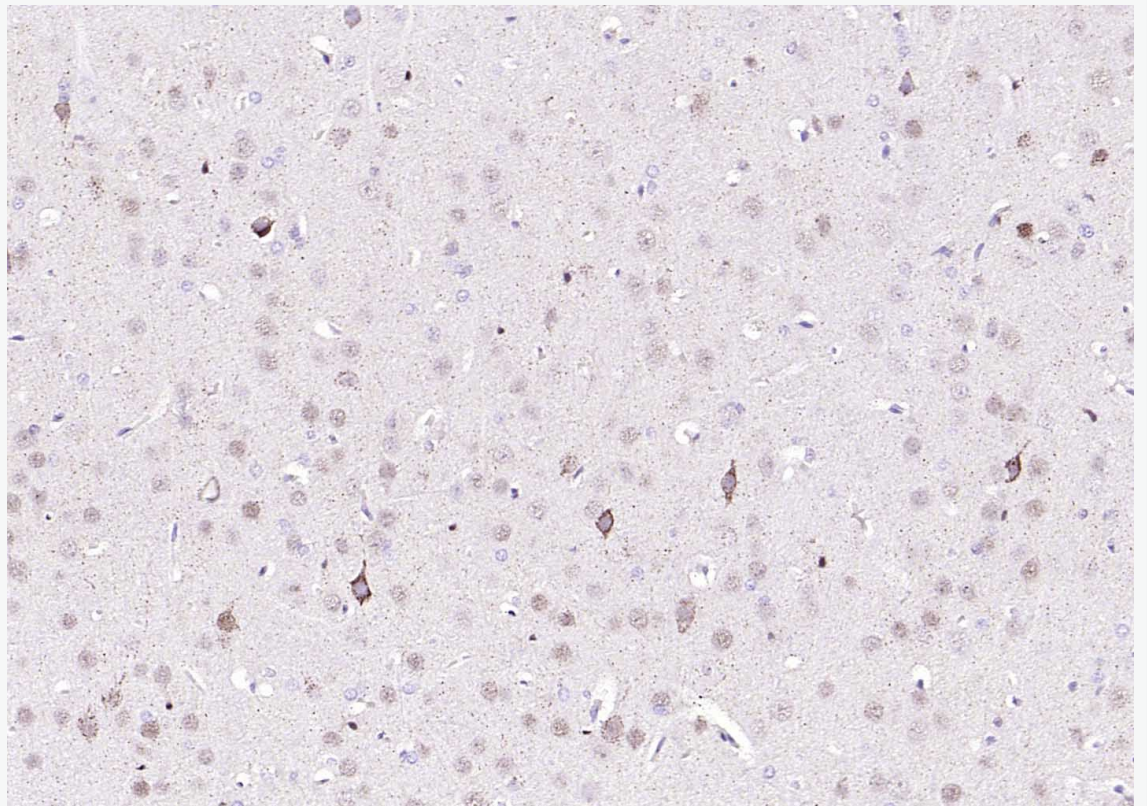
Paraformaldehyde-fixed, paraffin embedded (human myocardium); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Fumarase/FH) Monoclonal Antibody, Unconjugated (SLM-33167M) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Mouse)(sp-0024) instructions and DAB staining.



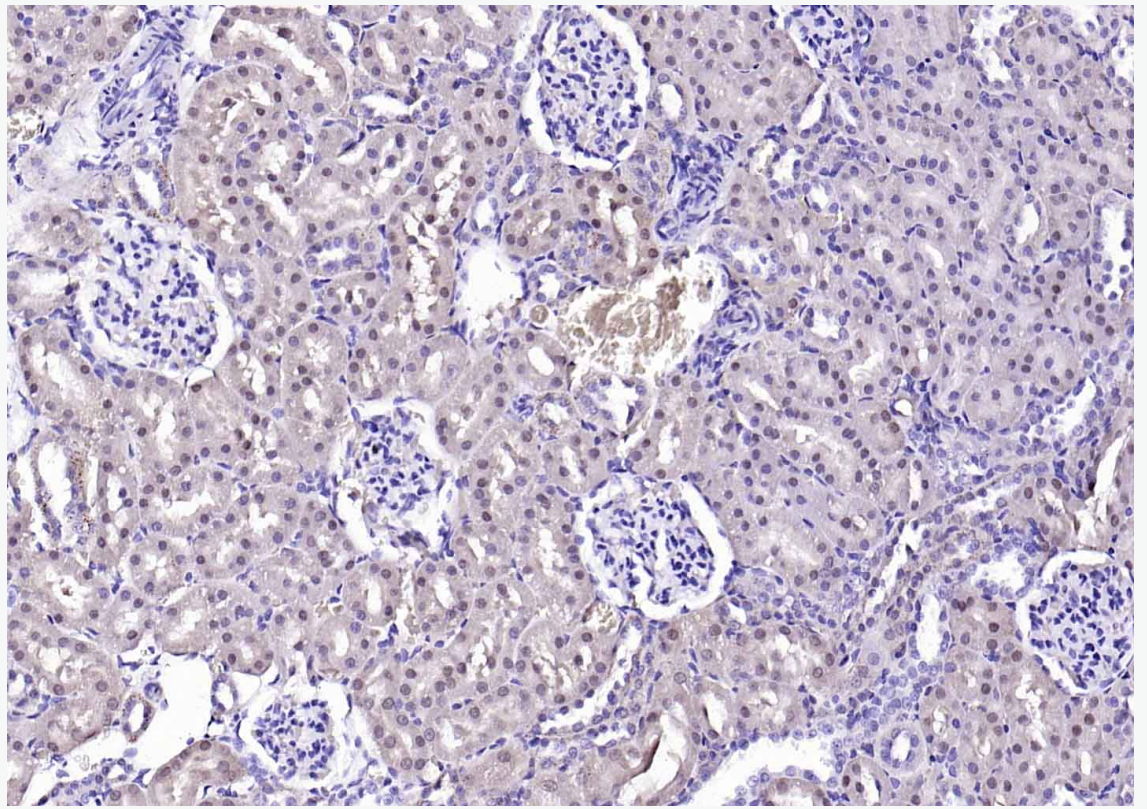
Paraformaldehyde-fixed, paraffin embedded (human gastric carcinoma); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Fumarase/FH) Monoclonal Antibody, Unconjugated (SLM-33167M) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Mouse)(sp-0024) instructions and DAB staining.



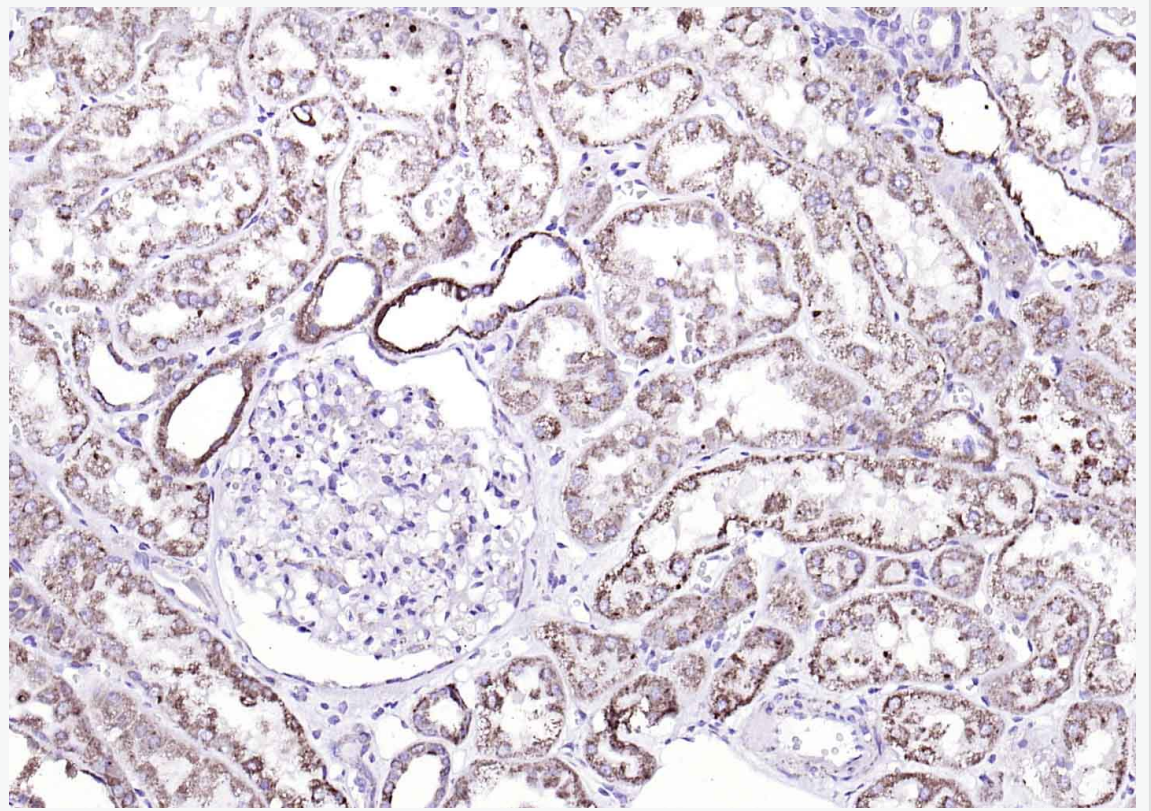
Paraformaldehyde-fixed, paraffin embedded (mouse heart); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Fumarase/FH) Monoclonal Antibody, Unconjugated (SLM-33167M) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Mouse)(sp-0024) instructions and DAB staining.



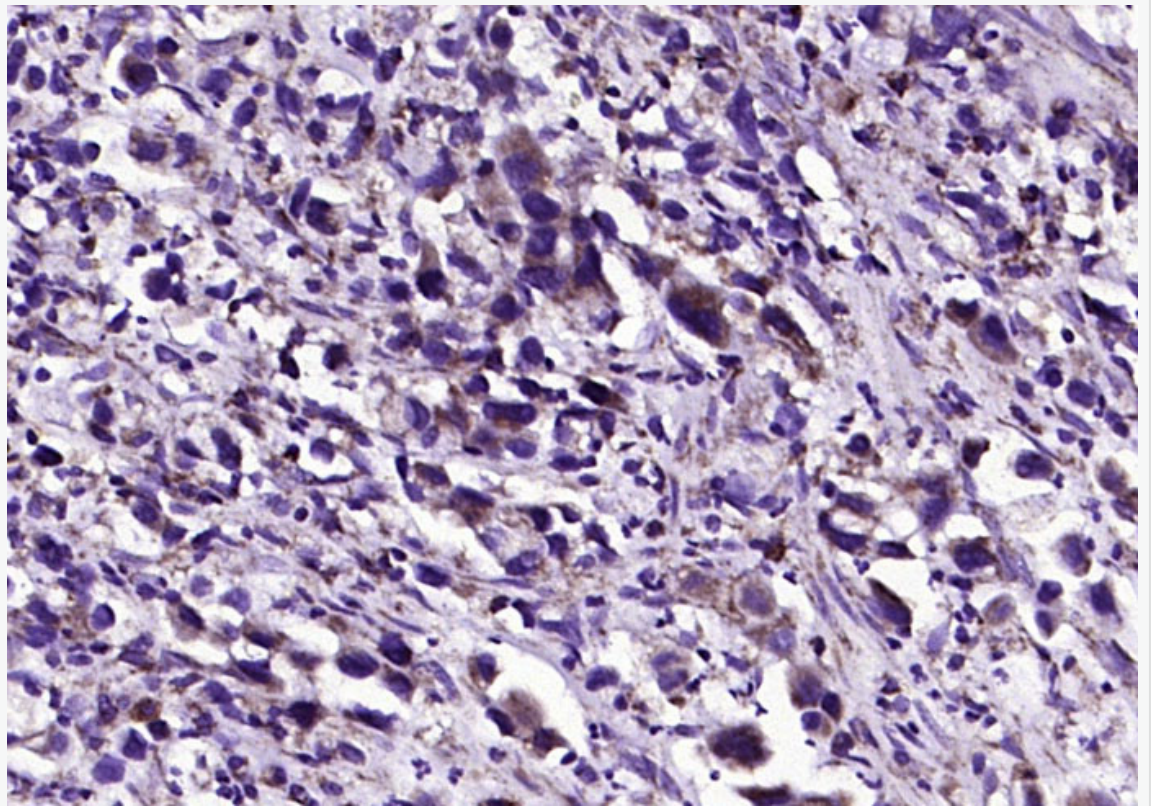
Paraformaldehyde-fixed, paraffin embedded (rat brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Fumarase/FH) Monoclonal Antibody, Unconjugated (SLM-33167M) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Mouse)(sp-0024) instructions and DAB staining.



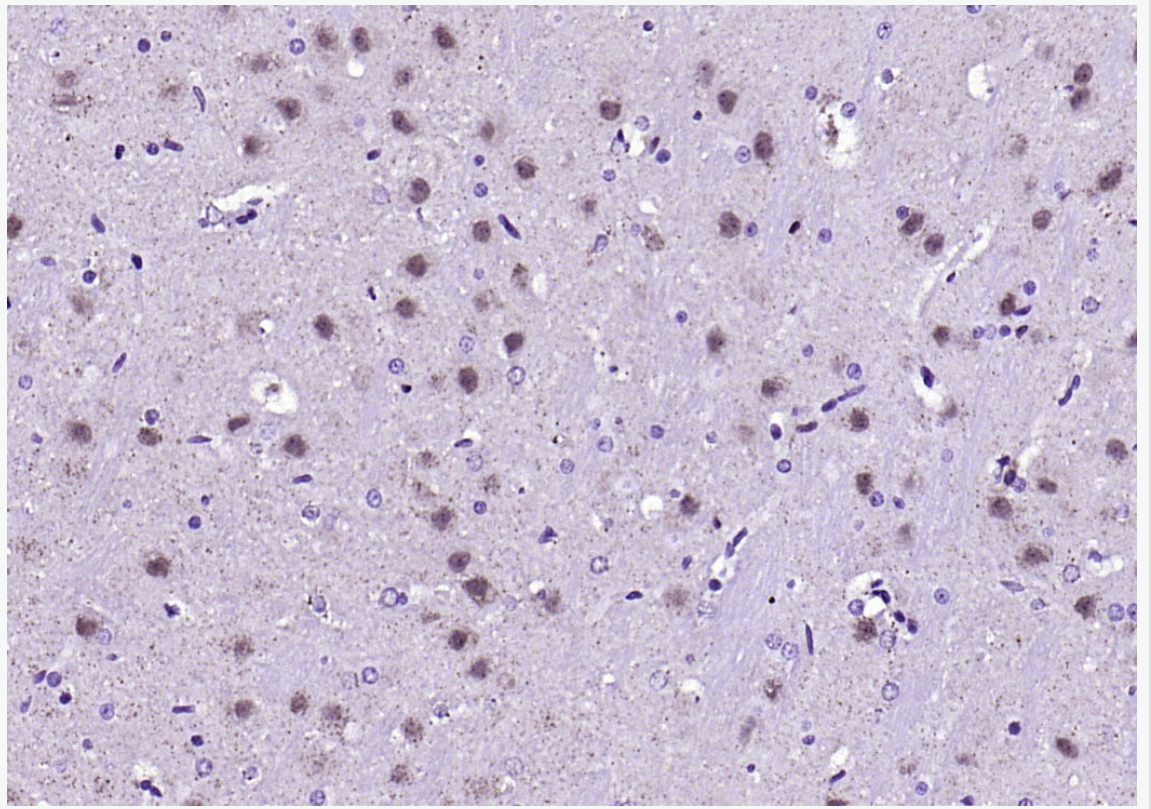
Paraformaldehyde-fixed, paraffin embedded (rat kidney); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Fumarase/FH) Monoclonal Antibody, Unconjugated (SLM-33167M) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Mouse)(sp-0024) instructions and DAB staining.



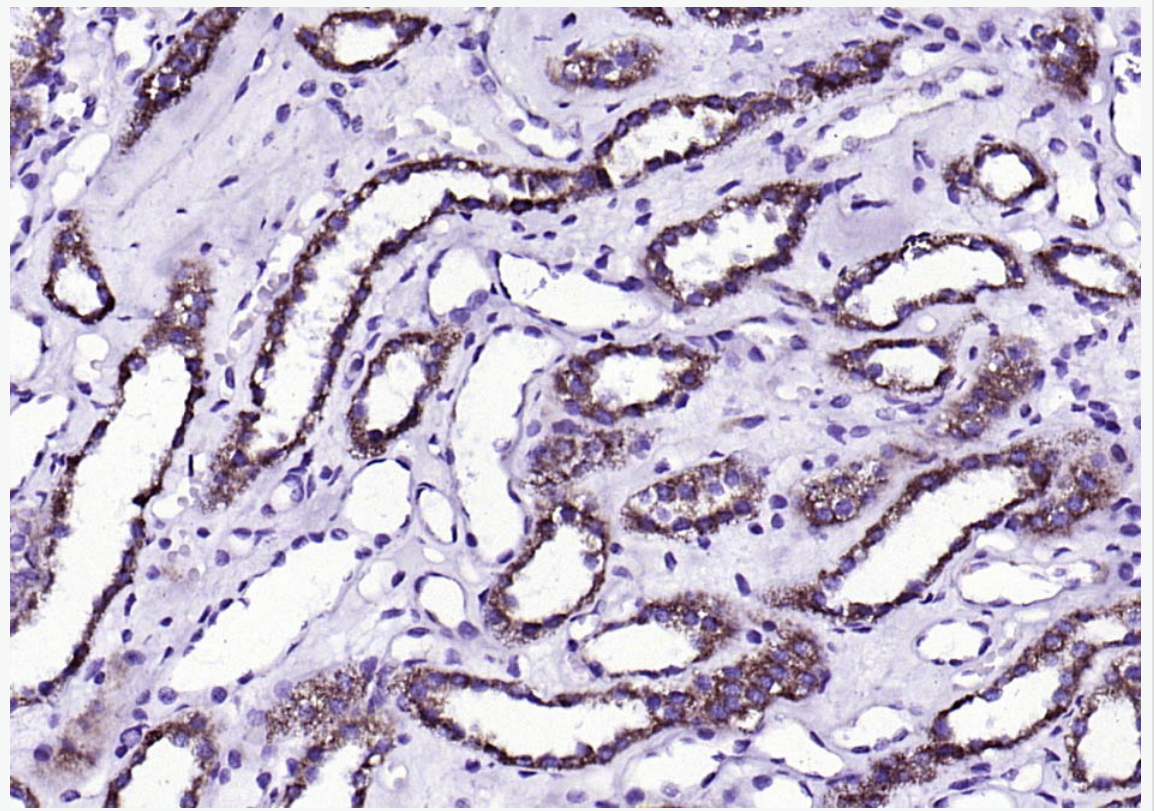
Paraformaldehyde-fixed, paraffin embedded (human kidney); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Fumarase/FH) Monoclonal Antibody, Unconjugated (SLM-33167M) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Mouse)(sp-0024) instructions and DAB staining.



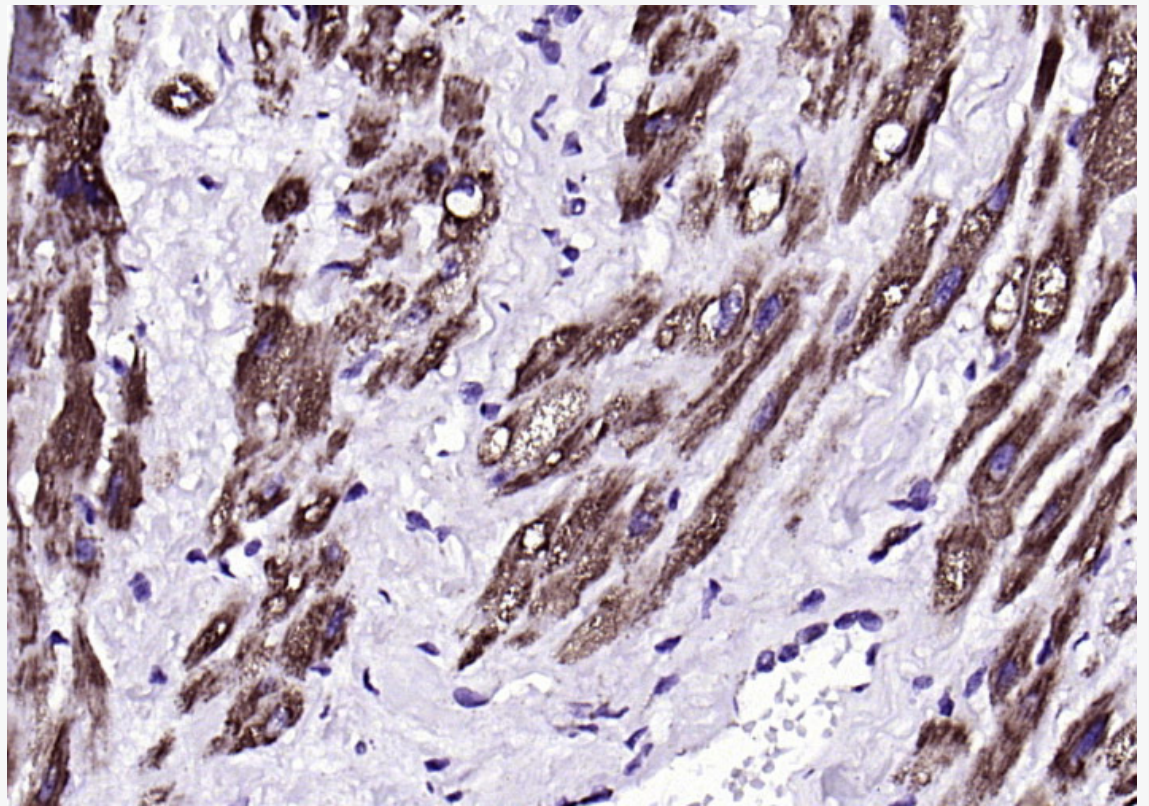
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Paraformaldehyde-fixed, paraffin embedded (human myocardium); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Fumarate/FH) Monoclonal Antibody, Unconjugated (SLM-33167M) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Mouse)(sp-0024) instructions and DAB staining.