

Fumarylacetoacetase polyclonal antibody

Catalog: BS67322

Host: Rabbit

Reactivity: Human

BackGround:

Fumarylacetoacetate hydrolase is the last enzyme in the tyrosine catabolism pathway. FAH deficiency is associated with Type 1 hereditary tyrosinemia (HT). This is an autosomal recessive inborn error of metabolism that occurs in both an acute and a chronic form. Clinical characteristics of the acute form include hepatic failure and death in infancy, whereas children with the chronic form have renal tubular dysfunction and hypophosphatemic rickets, progressive liver disease with development of hepatocellular carcinoma. Dietary treatment with restriction of tyrosine and phenylalanine alleviates the rickets, but liver transplantation has so far been the only definite treatment.

Product:

Liquid in 0.42% Potassium phosphate, 0.87% Sodium chloride, pH 7.3, 30% glycerol, and 0.01% sodium azide.

Molecular Weight:

~45 kDa

Swiss-Prot:

P16930

Purification&Purity:

The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen and the purity is > 95% (by SDS-PAGE).

Applications:

WB (1/500 - 1/2000), IHC (1/50 - 1/200), IF/ICC (1/10 - 1/100)

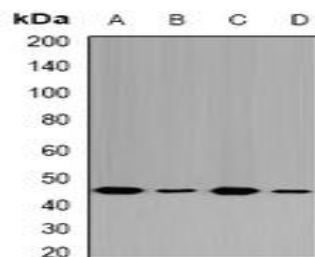
Storage&Stability:

Store at 4 °C short term. Aliquot and store at -20 °C long term. Avoid freeze-thaw cycles.

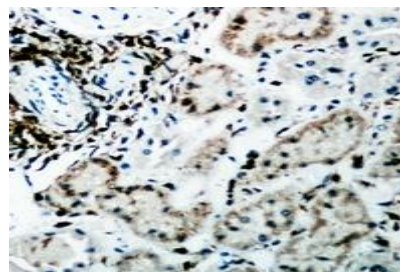
Specificity:

Recognizes endogenous levels of Fumarylacetoacetase protein.

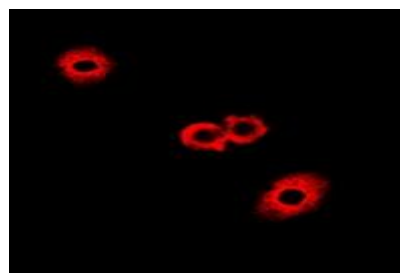
DATA:



Western blot analysis of Fumarylacetoacetase expression in HepG2 (A), MCF7 (B), mouse liver (C), rat kidney (D) whole cell lysates.



Immunohistochemical analysis of Fumarylacetoacetase staining in rat kidney formalin fixed paraffin embedded tissue section. The section was pre-treated using heat mediated antigen retrieval with sodium citrate buffer (pH 6.0). The section was then incubated with the antibody at room temperature and detected using an HRP conjugated compact polymer system. DAB was used as the chromogen. The section was then counterstained with haematoxylin and mounted with DPX.



Immunofluorescent analysis of Fumarylacetoacetase staining in A549 cells. Formalin-fixed cells were permeabilized with 0.1% Triton X-100 in TBS for 5-10 minutes and blocked with 3% BSA-PBS for 30 minutes at room temperature. Cells were probed with the primary antibody in 3% BSA-PBS and incubated overnight at 4 °C in a humidified chamber. Cells were washed with PBST and incubated with a DyLight

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PRODUCT DATA SHEET

Bioworld Technology, Inc.

594-conjugated secondary antibody (red) in PBS at room temperature in the dark.

Note:

For research use only, not for use in diagnostic procedure.

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