

FAH polyclonal antibody

Catalog: BS8270

Host: Rabbit

Reactivity: Human, Mouse, Rat

BackGround:

Fumarylacetoacetase is a 419 amino acid protein encoded by the human gene FAH. Fumarylacetoacetase catalyzes the hydrolysis of 4-fumarylacetoacetate, an intermediate in the metabolism of tyrosine, into acetoacetate and fumarate. Defects in FAH are the cause of tyrosinemia type I. It 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. Tyrosinemia type I is a rare condition, except in the Saguenay-lac-St-Jean region (province of Quebec, Canada) where the frequency is 1/1,846 newborns as the result of a founder effect.

Product:

1mg/ml in PBS with 0.1% Sodium Azide, 50% Glycerol.

Molecular Weight:

~ 46 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

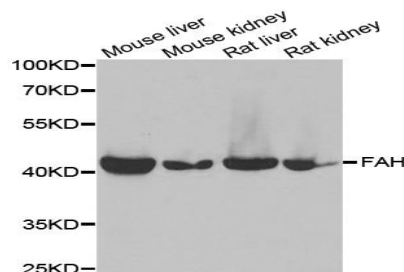
Storage&Stability:

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

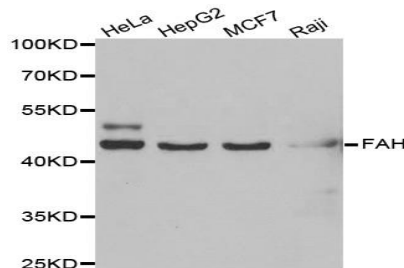
Specificity:

FAH polyclonal antibody detects endogenous levels of FAH protein.

DATA:



WesternBlot (WB) analysis of FAH polyclonal antibody



WesternBlot (WB) analysis of FAH polyclonal antibody

Note:

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

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