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Recombinant Human Fumarylacetoacetase/FAH Protein(C-6His)

产品货号: TP06128

第三版

别名:Fumarylacetoacetase; FAA; Beta-Diketonase; Fumarylacetoacetate Hydrolase; FAH

描述:Fumarylacetoacetase belongs to the FAH family. Fumarylacetoacetase is primary expressed in liver and kidney. It exists as a homodimer and catalyzes the hydrolysis of 4-fumarylacetoacetate into fumarate and acetoacetate. Defects in Fumarylacetoacetase cause tyrosinemia type 1, which is congenital metabolism defect characterized by elevated levels of tyrosine in the blood and urine, and hepatorenal manifestations. Typical features include renal tubular injury, self-mutilation, hepatic necrosis, episodic weakness, and seizures.

配方:PBS

分子量:47.4 kDa

序列:Ser2-Ser419

纯度:> 95% by HPLC

浓度:

内毒素:<1.0 EU per 1 ug of protein (determined by LAL method)

存储: +4 ° C 保存 (1-2 周). 长期保存在-20 ° C或者-70 ° C. 避免反复冻融.