

## Recombinant Human Fumarylacetoacetase/FAH Protein (His Tag)

**Catalog Number:** PKSH032463

**Note:** Centrifuge before opening to ensure complete recovery of vial contents.

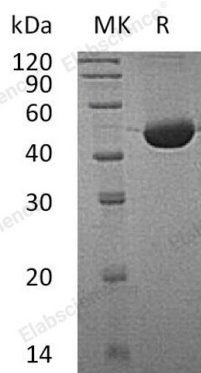
### Description

<b>Species</b>	Human
<b>Source</b>	HEK293 Cells-derived Human Fumarylacetoacetase;FAH protein Ser2-Ser419, with an C-terminal His
<b>Calculated MW</b>	47.4 kDa
<b>Observed MW</b>	43 kDa
<b>Accession</b>	P16930
<b>Bio-activity</b>	Not validated for activity

### Properties

<b>Purity</b>	> 95 % as determined by reducing SDS-PAGE.
<b>Endotoxin</b>	< 1.0 EU per µg of the protein as determined by the LAL method.
<b>Storage</b>	Generally, lyophilized proteins are stable for up to 12 months when stored at -20 to -80 °C. Reconstituted protein solution can be stored at 4-8°C for 2-7 days. Aliquots of reconstituted samples are stable at < -20°C for 3 months.
<b>Shipping</b>	This product is provided as lyophilized powder which is shipped with ice packs.
<b>Formulation</b>	Lyophilized from a 0.2 µm filtered solution of 20mM Tris-HCl, 150mM NaCl, pH 8.5. Normally 5% - 8% trehalose, mannitol and 0.01% Tween 80 are added as protectants before lyophilization.
	Please refer to the specific buffer information in the printed manual.
<b>Reconstitution</b>	Please refer to the printed manual for detailed information.

### Data



> 95 % as determined by reducing SDS-PAGE.

### Background

Fumarylacetoacetase belongs to the FAH family. Fumarylacetoacetase is primarily 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 a 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.

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