



Recombinant Protein Technical Manual

**Recombinant Human Fumarylacetoacetase/FAH
Protein (His Tag)**
RPES4423

Product Data:

Product SKU: RPES4423

Size: 10µg

Species: Human

Expression host: Human Cells

Uniprot: P16930

Protein Information:

Molecular Mass: 47.4 kDa

AP Molecular Mass: 47 kDa

Tag: C-6His

Bio-activity:

Purity: > 95 % as determined by reducing SDS-PAGE.

Endotoxin: < 1.0 EU per µg as determined by the LAL method.

Storage: 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.

Shipping: This product is provided as lyophilized powder which is shipped with ice packs.

Formulation: Lyophilized from a 0.2 µm filtered solution of PBS, pH7.4.

Reconstitution: Please refer to the printed manual for detailed information.

Application:

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

Immunogen Information:

Sequence: Ser2-Ser419

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.