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Recombinant Human Fumarate Hydratase / FH Protein (His tag)

产品货号: TP06800

第三版

别名:FMRD;HLRCC;LRCC;MCL;MCUL1

**描述:**Fumarate Hydratase (FH) is an enzymatic component of the tricarboxylic acid (TCA) cycle, or Krebs cycle, and catalyzes the formation of L-malate from fumarate. It exists in both a cytosolic form and an N-terminal extended form, differing only in the translation start site used. The N-terminal extended form is targeted to the mitochondrion, where the removal of the extension generates the same form as in the cytoplasm. Fumarate Hydratase is similar to some thermostable class II fumarases and functions as a homotetramer. Mutations in this gene can cause fumarase deficiency and lead to progressive encephalopathy. Individuals with hemizygous germline fumarate hydratase (FH) mutations are predisposed to renal cancer. These tumors predominantly exhibit functional inactivation of the remaining wild-type allele, implicating FH inactivation as a tumor-promoting event.

**配方:**PBS

**分子量:**52 kDa

**序列:**Ala 45-Lys 510

**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.