

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human Ketohexokinase in direct ELISAs.
Source	Monoclonal Mouse IgG ₁ Clone # 1020613
Purification	Protein A or G purified from cell culture supernatant
Immunogen	<i>E. coli</i> -derived human Ketohexokinase Met1-Val298 Accession # P50053
Conjugate	Alexa Fluor 647 Excitation Wavelength: 650 nm Emission Wavelength: 668 nm
Formulation	Supplied 0.2mg/ml in 1X PBS with RDF1 and 0.09% Sodium Azide
*Contains <0.1% Sodium Azide, which is not hazardous at this concentration according to GHS classifications. Refer to the Safety Data Sheet (SDS) for additional information and handling instructions.	

APPLICATIONS

Please Note: Optimal dilutions should be determined by each laboratory for each application. [General Protocols](#) are available in the Technical Information section on our website.

Western Blot	Optimal dilution of this antibody should be experimentally determined.
Immunohistochemistry	Optimal dilution of this antibody should be experimentally determined.

PREPARATION AND STORAGE

Shipping	The product is shipped with polar packs. Upon receipt, store it immediately at the temperature recommended below.
Stability & Storage	Protect from light. Do not freeze. 12 months from date of receipt, 2 to 8 °C as supplied

BACKGROUND

KHK1(Ketohexokinase) catalyzes conversion of fructose to fructose-1-phosphate (1). It is the first enzyme that catabolizes dietary fructose. Mutation of this protein is the molecular basis for essential fructosuria, a clinically benign condition characterized by the incomplete metabolism of fructose in the liver, leading to its excretion in urine (2, 3). Essential fructosuria does not have any clinical manifestations and no treatment is required. However, deficiency of aldolase B, the second enzyme involved in the metabolism of fructose results in the accumulation of fructose-1-phosphate in the blood, which causes fructosemia or hereditary fructose intolerance (4). High level of fructose-1-phosphate inhibits the production of glucose and results in diminished regeneration of adenosine triphosphate. Patients with fructosemia have symptoms of elevated uric acid, growth abnormalities, and coma if untreated. Therefore, inhibition of KHK1 may lead to a cure for fructosemia. High level of expression of KHK1 is found in liver, kidney, gut, spleen and pancreas. Low levels of expression of KHK1 is found in heart, muscle, brain, and eye (3). The enzymatic activity of recombinant human KHK1 is measured using a phosphatase-coupled method (5).

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