

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 750 Excitation Wavelength: 749 nm Emission Wavelength: 775 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).

PRODUCT SPECIFIC NOTICES

This product is provided under an agreement between Life Technologies Corporation and R&D Systems, Inc, and the manufacture, use, sale or import of this product is subject to one or more US patents and corresponding non-US equivalents, owned by Life Technologies Corporation and its affiliates. The purchase of this product conveys to the buyer the non-transferable right to use the purchased amount of the product and components of the product only in research conducted by the buyer (whether the buyer is an academic or for-profit entity). The sale of this product is expressly conditioned on the buyer not using the product or its components (1) in manufacturing; (2) to provide a service, information, or data to an unaffiliated third party for payment; (3) for therapeutic, diagnostic or prophylactic purposes; (4) to resell, sell, or otherwise transfer this product or its components to any third party, or for any other commercial purpose. Life Technologies Corporation will not assert a claim against the buyer of the infringement of the above patents based on the manufacture, use or sale of a commercial product developed in research by the buyer in which this product or its components was employed, provided that neither this product nor any of its components was used in the manufacture of such product. For information on purchasing a license to this product for purposes other than research, contact Life Technologies Corporation, Cell Analysis Business Unit, Business Development, 29851 Willow Creek Road, Eugene, OR 97402, Tel: (541) 465-8300. Fax: (541) 335-0354.