

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human Ketohehexokinase 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 Ketohehexokinase Met1-Val298 Accession # P50053
Formulation	Lyophilized from a 0.2 µm filtered solution in PBS with Trehalose. See Certificate of Analysis for details. *Small pack size (-SP) is supplied either lyophilized or as a 0.2 µm filtered solution in PBS.

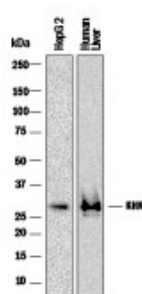
APPLICATIONS

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.

	Recommended Concentration	Sample
Western Blot	2 µg/mL	See Below
Immunohistochemistry	5-25 µg/mL	See Below

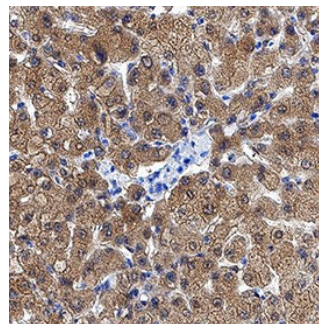
DATA

Western Blot



Detection of Human Ketohehexokinase by Western Blot. Western blot shows lysates of HepG2 human hepatocellular carcinoma cell line and human liver tissue. PVDF membrane was probed with 2 µg/mL of Mouse Anti-Human Ketohehexokinase Monoclonal Antibody (Catalog # MAB8177) followed by HRP-conjugated Anti-Mouse IgG Secondary Antibody (Catalog # HAF018). A specific band was detected for Ketohehexokinase at approximately 30 kDa (as indicated). This experiment was conducted under reducing conditions and using Immunoblot Buffer Group 1.

Immunohistochemistry



Ketohehexokinase in Human Liver. Ketohehexokinase was detected in immersion fixed paraffin-embedded sections of human liver using Mouse Anti-Human Ketohehexokinase Monoclonal Antibody (Catalog # MAB8177) at 5 µg/mL for 1 hour at room temperature followed by incubation with the Anti-Mouse IgG VisUCyte™ HRP Polymer Antibody (Catalog # VC001). Before incubation with the primary antibody, tissue was subjected to heat-induced epitope retrieval using Antigen Retrieval Reagent-Basic (Catalog # CTS013). Tissue was stained using DAB (brown) and counterstained with hematoxylin (blue). Specific staining was localized to cytoplasm in hepatocytes. View our protocol for [IHC Staining with VisUCyte HRP Polymer Detection Reagents](#).

PREPARATION AND STORAGE

Reconstitution	Reconstitute at 0.5 mg/mL in sterile PBS.
Shipping	The product is shipped at ambient temperature. Upon receipt, store it immediately at the temperature recommended below. *Small pack size (-SP) is shipped with polar packs. Upon receipt, store it immediately at -20 to -70 °C
Stability & Storage	Use a manual defrost freezer and avoid repeated freeze-thaw cycles. <ul style="list-style-type: none"> 12 months from date of receipt, -20 to -70 °C as supplied. 1 month, 2 to 8 °C under sterile conditions after reconstitution. 6 months, -20 to -70 °C under sterile conditions after reconstitution.

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).

References:

1. Trinh, C.H. et al. (2009) Acta. Crystallogr. D Biol Crystallogr. 65:201.
2. Zhang, X. et al. (2011) Bioorg. Med. Chem. Lett. 21:4762.
3. Bonthron, D.T. et al. (1994) Hum. Mol. Genet. 3:1627.
4. Kaiser, U.B. and Hegele, R. A. (1991). Am. J. Med. Sci. 302: 364.
5. Wu, Z.L. (2011) PLoS One 6:e23172.