KHK dimer phosphorylates Fru to Fru 1-P

Cameselle, JC., D'Eustachio, P., Hill, DP., Jassal, B., Ribeiro, JM.

European Bioinformatics Institute, New York University Langone Medical Center, Ontario Institute for Cancer Research, Oregon Health and Science University.

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Introduction

Reactome is open-source, open access, manually curated and peer-reviewed pathway database. Pathway annotations are authored by expert biologists, in collaboration with Reactome editorial staff and cross-referenced to many bioinformatics databases. A system of evidence tracking ensures that all assertions are backed up by the primary literature. Reactome is used by clinicians, geneticists, genomics researchers, and molecular biologists to interpret the results of high-throughput experimental studies, by bioinformaticians seeking to develop novel algorithms for mining knowledge from genomic studies, and by systems biologists building predictive models of normal and disease variant pathways.

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Literature references

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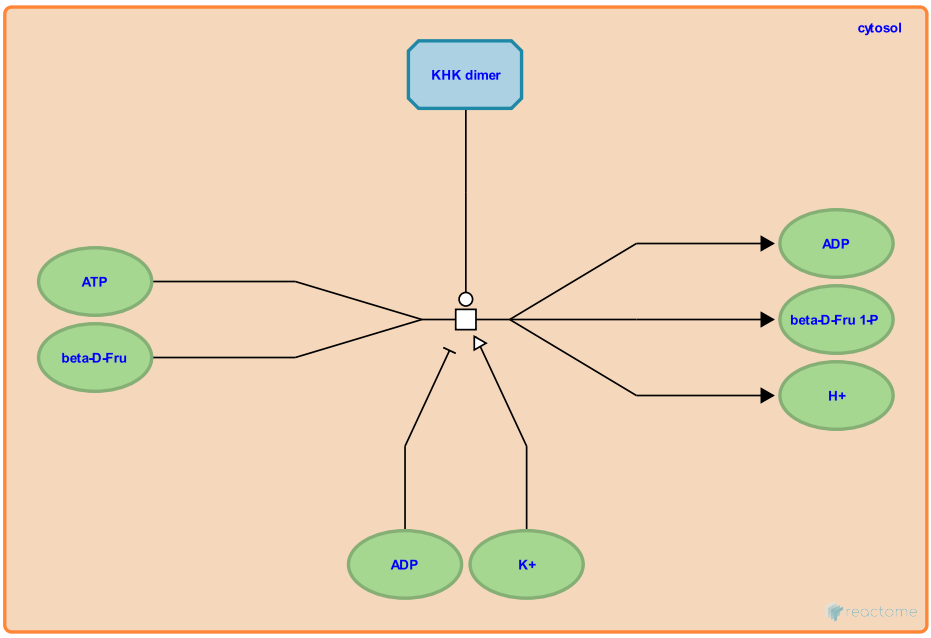
Stable identifier: R-HSA-70333

Type: transition

Compartments: cytosol



Cytosolic ketohexokinase (KHK, also known as fructokinase) catalyzes the reaction of D-fructose (Fru) and ATP to form D-fructose 1-phosphate (Fru 1-P) and ADP. Two isoforms of the enzyme, A and C, are encoded by alternatively spliced forms of the gene; both form catalytically active dimers. The C isoform is predominant in liver and kidney tissues, has high affinity for fructose, and is probably responsible for the bulk of fructose phosphorylation in vivo (Asipu et al. 2003; Trinh et al. 2009). The A isoform is found in lower levels in many other tissues and may serve a role in fructose metabolism outside of liver and kidney (Funari et al. 2005). The physiological role of KHK has been established from metabolic and DNA sequencing studies of patients with essential fructosuria (Bonthron et al. 1994) and in mouse models for this disease (Diggle et al. 2010; Ishimoto et al. 2012).

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