

# **GFPT1** Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab) Catalog # AP55140

### **Product Information**

Application Primary Accession Reactivity Host Clonality Calculated MW Physical State Immunogen Epitope Specificity Isotype Purity	WB, IHC-P, IHC-F, IF, ICC, E Q06210 Rat, Pig, Dog Rabbit Polyclonal 78806 Liquid KLH conjugated synthetic peptide derived from human GFPT1 601-699/699 IgG affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 1 glutamine amidotransferase type-2 domain. Contains 2 SIS
SUBUNIT	Homotetramer
DISEASE	Defects in GFPT1 are the cause of limb-girdle myasthenia with tubular aggregates (LGMTA) [MIM:610542]. A congenital myasthenic syndrome characterized by onset of proximal muscle weakness in the first decade. Individuals with this condition have a recognizable pattern of weakness of shoulder and pelvic girdle muscles, and sparing of ocular or facial muscles. EMG classically shows a decremental response to repeated nerve stimulation, a sign of neuromuscular junction dysfunction. Affected individuals show a favorable response to acetylcholinesterase (AChE) inhibitors.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Glutamine:fructose-6-phosphate amidotransferase (GFAT1) is the first and rate-limiting enzyme for the entry of glucose into the hexosamine biosynthesis pathway (HBP) in mammals. GFAT1, a member of the N-terminal nucleophile class of amidotransferases, converts fructose-6-phosphate into N-acetylglucosamine-6-phosphate. Hyperglycemia-induced insulin resistance, a condition in which exposure to high concentrations of glucose and insulin results in insulin resistance, may result from increased glucose metabolism through the HBP. Hypergylcemia-induced insulin resistance is a characteristic feature of type 2 diabetes. Consequently, GFAT1 is a potential therapeutic target in the treatment of type 2 diabetes.

## **Additional Information**

Gene ID	2673
Other Names	Glutaminefructose-6-phosphate aminotransferase [isomerizing] 1, 2.6.1.16,

	D-fructose-6-phosphate amidotransferase 1, Glutamine:fructose-6-phosphate amidotransferase 1, GFAT 1, GFAT1, Hexosephosphate aminotransferase 1, GFPT1, GFAT, GFPT
Target/Specificity	Isoform 1 is predominantly expressed in skeletal muscle. Not expressed in brain. Seems to be selectively expressed in striated muscle.
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC=1:100-500,IF=1:100-50 0,ELISA=1:5000-10000
Format	0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

#### **Protein Information**

Name	GFPT1
Synonyms	GFAT, GFPT
Function	Controls the flux of glucose into the hexosamine pathway. Most likely involved in regulating the availability of precursors for N- and O-linked glycosylation of proteins. Regulates the circadian expression of clock genes BMAL1 and CRY1 (By similarity). Has a role in fine tuning the metabolic fluctuations of cytosolic UDP-GlcNAc and its effects on hyaluronan synthesis that occur during tissue remodeling (PubMed: <u>26887390</u> ).
Tissue Location	Isoform 1 is predominantly expressed in skeletal muscle. Not expressed in brain. Seems to be selectively expressed in striated muscle.

#### Images



Tissue/cell: human lung carcinoma; 4% Paraformaldehyde-fixed and paraffin-embedded; Antigen retrieval: citrate buffer ( 0.01M, pH 6.0 ), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37°C for 20 min; Incubation: Anti-GFPT1 Polyclonal Antibody, Unconjugated(AP55140) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining

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