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GFPT1 Rabbit pAb

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Catalog # AP55140

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q06210
Reactivity	Human
Predicted	Rat, Pig, Dog, Mouse, Rabbit, Chicken, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	78806
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human GFPT1
Epitope Specificity	601-699/699
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
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	Glutamine--fructose-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 (HGNC:4241), GFAT, GFPT
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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.

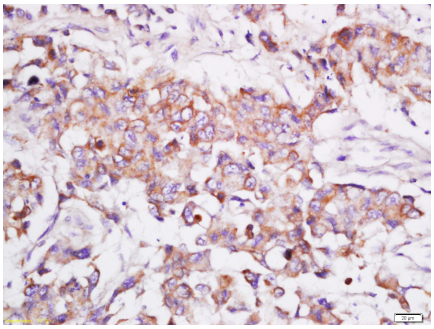
Protein Information

Name	GFPT1 (HGNC:4241)
Synonyms	GFAT, GFPT
Function	Rate-limiting enzyme of the hexosamine biosynthetic pathway (HBP) that catalyzes the formation of glucosamine-6-phosphate from fructose-6-phosphate and glutamine, thereby controlling the flux of glucose into this pathway (PubMed:32019926, PubMed:35229715). Inhibited by UDP-N-acetylglucosamine (UDP-GlcNAc) through a feedback loop (PubMed:32019926, PubMed:35229715). Fine-tunes the metabolic fluctuations of UDP-GlcNAc and its impacts on hyaluronan synthesis during tissue remodeling (PubMed:26887390). Via control of the HPB, regulates the availability of precursors for N- and O-linked protein glycosylation and modulates peripheral clock oscillation (By similarity).
Tissue Location	[Isoform 1]: Predominantly expressed in skeletal muscle. Not expressed in brain. Seems to be selectively expressed in striated muscle.

Background

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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℃ 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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