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AGL Antibody (N-term)

Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP2402c

Product Information

Application	IF, WB, E
Primary Accession	P35573
Reactivity	Human
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB5106
Calculated MW	174764

Additional Information

Gene ID	178
Other Names	Glycogen debranching enzyme, Glycogen debrancher, 4-alpha-glucanotransferase, Oligo-1, 4-1, 4-glucantransferase, Amylo-alpha-1, 6-glucosidase, Amylo-1, 6-glucosidase, Dextrin 6-alpha-D-glucosidase, AGL, GDE
Target/Specificity	This AGL antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide selected from the N-terminal region of human AGL.
Dilution	IF~~1:10~50 WB~~1:1000 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is prepared by Saturated Ammonium Sulfate (SAS) precipitation followed by dialysis against PBS.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	AGL Antibody (N-term) is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	AGL
Synonyms	GDE
Function	Multifunctional enzyme acting as 1,4-alpha-D-glucan:1,4- alpha-D-glucan

	4-alpha-D-glycosyltransferase and amylo-1,6-glucosidase in glycogen degradation.
Cellular Location	Cytoplasm. Note=Under glycogenolytic conditions localizes to the nucleus
Tissue Location	Liver, kidney and lymphoblastoid cells express predominantly isoform 1; whereas muscle and heart express not only isoform 1, but also muscle-specific isoform mRNAs (isoforms 2, 3 and 4). Isoforms 5 and 6 are present in both liver and muscle

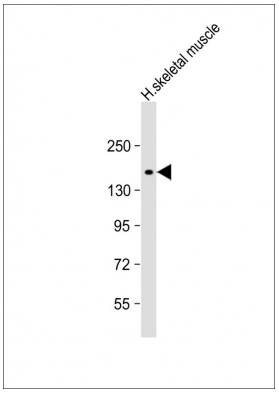
Background

AGL is a glycogen debrancher enzyme which is involved in glycogen degradation. This enzyme has two independent catalytic activities which occur at different sites on the protein: a 4-alpha-glucotransferase activity and a amylo-1,6-glucosidase activity. Mutations in the AGL gene are associated with glycogen storage disease although a wide range of enzymatic and clinical variability occurs which may be due to tissue-specific alternative splicing.

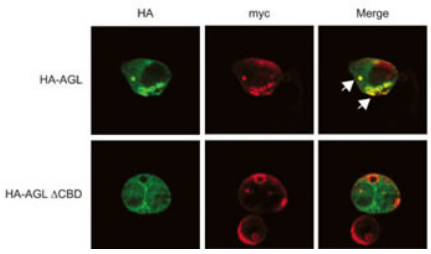
References

Horinishi, A., et al., J. Hum. Genet. 47(2):55-59 (2002).
Shen, J., et al., Hum. Mutat. 9(1):37-40 (1997).
Bao, Y., et al., Genomics 38(2):155-165 (1996).
Shen, J., et al., J. Clin. Invest. 98(2):352-357 (1996).
Yang, B.Z., et al., J. Biol. Chem. 267(13):9294-9299 (1992).

Images



Anti-AGL Antibody (M15) at 1:1000 dilution + human skeletal muscle lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size : 175 kDa Blocking/Dilution buffer: 5% NFDM/TBST.



Expression of myc-GS causes wild type but not the □□CBD mutant of AGL to aggregate around the PAS-stain-positive inclusions. HepG2 cells were transfected with either HA-tagged wild-type AGL (HA-AGL) or HA-AGL □□CBD. Cells were fixed in formalin and processed for IF using anti-HA (green) and anti-myc (red) antibodies. White arrows indicate colocalization of HA-AGL and myc-GS.