10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

AGL Antibody (C-term)

Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP2402B

Product Information

Application	WB, IF, E
Primary Accession	P35573
Reactivity	Human
Predicted	Mouse, Rat, Bovine, Canine, Rabbit, Chicken
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Calculated MW	174764
Antigen Region	1479-1510

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 between 1479-1510 amino acids from the C-terminal region of human AGL.
Dilution	WB~~1:1000 IF~~1:10~50 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 (C-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

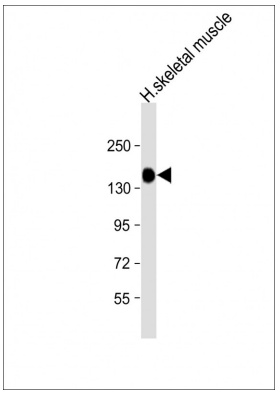
Background

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.

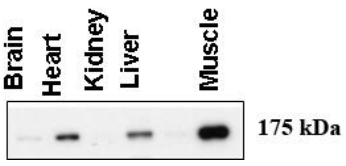
References

Horinishi, A., et al., J. Hum. Genet. 47(2):55-59 (2002).
Bao, Y., et al., Genomics 38(2):155-165 (1996).
Yang, B.Z., et al., J. Biol. Chem. 267(13):9294-9299 (1992).
Yang-Feng, T.L., et al., Genomics 13(4):931-934 (1992).
Bao, Y., et al., Gene 197 (1-2), 389-398 (1997).

Images

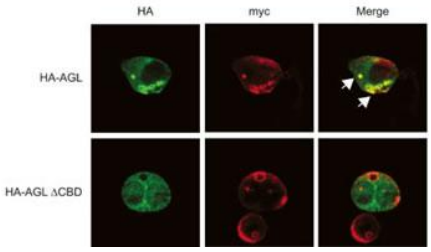


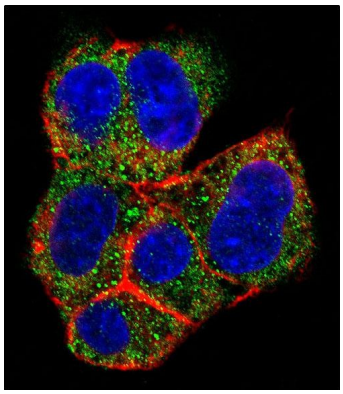
Anti-AGL Antibody (C-term) at 1:8000 dilution + human skeletal muscle lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size : 175 kDa Blocking/Dilution buffer: 5% NFDM/TBST.



Western blot using anti-AGL (C-term) antibody (AP2402c) at 1:1000 dilution. A total of 20 ug of lysates was loaded for each tissue. Data courtesy of Dr. Alan Cheng, Department of Internal Medicine, Life Sciences Institute, University of Michigan Medical Center, Ann Arbor, Michigan.

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.





Confocal immunofluorescent analysis of AGL Antibody (C-term)(Cat#AP2402b) with HepG2 cell followed by Alexa Fluor 488-conjugated goat anti-rabbit lgG (green). Actin filaments have been labeled with Alexa Fluor 555 phalloidin (red).DAPI was used to stain the cell nuclear (blue).

Citations

- DePaoli-Roach AA., et al. Genetic depletion of the malin E3 ubiquitin ligase in mice leads to lafora bodies and the accumulation of insoluble laforin.J Biol Chem. 2010 Aug 13;285(33):25372-81. doi: 10.1074/jbc.M110.148668.
- Fast-twitch sarcomeric and glycolytic enzyme protein loss in inclusion body myositis.
- A role for AGL ubiquitination in the glycogen storage disorders of Lafora and Cori's disease.
- Laforin-Malin Complex Degrades Polyglucosan Bodies in Concert with Glycogen Debranching Enzyme and Brain Isoform Glycogen Phosphorylase.
- Anti-retinal antibodies in patients with macular telangiectasia type 2.
- Loss of Glycogen Debranching Enzyme AGL Drives Bladder Tumor Growth via Induction of Hyaluronic Acid Synthesis.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.