

AGL Antibody (C-term)

Purified Rabbit Polyclonal Antibody (Pab) Catalog # AP2402B

Specification

AGL Antibody (C-term) - Product Information

Application IF, WB,E
Primary Accession P35573
Reactivity Human
Host Rabbit
Clonality Polyclonal
Isotype Rabbit IgG
Antigen Region 1479-1510

AGL Antibody (C-term) - 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

IF~~1:10~50 WB~~1:8000

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.

AGL Antibody (C-term) - 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

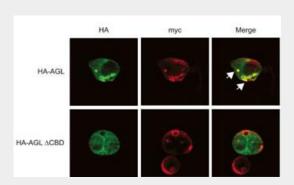
Liver, kidney and lymphoblastoid cells express predominantly isoform 1; whereas muscle and heart express not only isoform 1, but also muscle-specific isoform mRNAs (isoforms 2, 3 and 4). Isoforms 5 and 6 are present in both liver and muscle

AGL Antibody (C-term) - Protocols

Provided below are standard protocols that you may find useful for product applications.

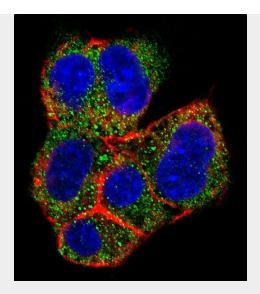
- Western Blot
- Blocking Peptides
- Dot Blot
- <u>Immunohistochemistry</u>
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

AGL Antibody (C-term) - Images

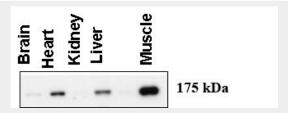


Expression of myc-GS causes wild type but not the $\tilde{A} \square CBD$ mutant of AGL to aggregate around the PAS-stain-positive inclusions. HepG2 cells were transfected with either HA-tagged wild-type AGL (HA-AGL) or HA-AGL $\tilde{A} \square CBD$. Cells were fixed in formalin and processed for IF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

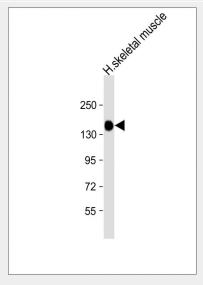




Confocal immunofluorescent analysis of AGL Antibody (C-term)(Cat#AP2402b) with HepG2 cell followed by Alexa Fluor 488-conjugated goat anti-rabbit IgG (green). Actin filaments have been labeled with Alexa Fluor 555 phalloidin (red).DAPI was used to stain the cell nuclear (blue).



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.



Anti-AGL Antibody (C-term) at 1:8000 dilution + human skeletal muscle lysate Lysates/proteins at 20 μ g 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.

AGL Antibody (C-term) - 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



Tel: 858.875.1900 Fax: 858.875.1999

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.

AGL Antibody (C-term) - 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).

AGL Antibody (C-term) - Citations

- Loss of Glycogen Debranching Enzyme AGL Drives Bladder Tumor Growth via Induction of Hyaluronic Acid Synthesis.
- 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.
- 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. I 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.