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

AGL Antibody (Center)

Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP2402a

Product Information

Application	WB, IF, E
Primary Accession	P35573
Reactivity	Human
Predicted	Mouse, Rat, Bovine, Canine, Rabbit
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Calculated MW	174764
Antigen Region	357-387

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 357-387 amino acids from the Central 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 (Center) 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

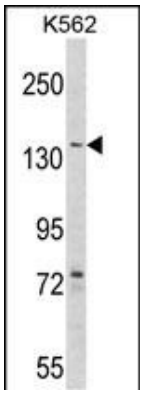
Background

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.

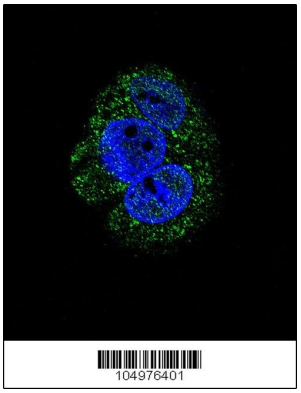
References

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



Western blot analysis of hAGL-C371 (Cat. #AP2402a) in K562 cell line lysates (35ug/lane). AGL (arrow) was detected using the purified Pab.



Confocal immunofluorescent analysis of AGL Antibody (Center)(Cat#AP2402a) with HepG2 cell followed by Alexa Fluor 488-conjugated goat anti-rabbit lgG (green). DAPI was used to stain the cell nuclear (blue).

Citations

- Fast-twitch sarcomeric and glycolytic enzyme protein loss in inclusion body myositis.
- Abnormal metabolism of glycogen phosphate as a cause for Lafora disease.
- A role for AGL ubiquitination in the glycogen storage disorders of Lafora and Cori\'s disease.
- Genetic depletion of the malin E3 ubiquitin ligase in mice leads to lafora bodies and the accumulation of insoluble laforin.
- Muscle glycogen remodeling and glycogen phosphate metabolism following exhaustive exercise of wild type and laforin knockout mice.
- 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'.