

## **AGL Antibody (Center)**

Purified Rabbit Polyclonal Antibody (Pab) Catalog # AP2402a

## **Specification**

## **AGL Antibody (Center) - Product Information**

Application WB, IF,E
Primary Accession P35573
Reactivity Human
Host Rabbit
Clonality Polyclonal
Isotype Rabbit IgG
Antigen Region 357-387

# **AGL Antibody (Center) - 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.

# **AGL Antibody (Center) - 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**

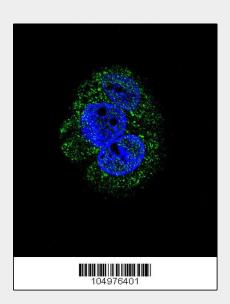
Liver, kidney and lymphoblastoid cells express predominantly isoform 1; whereas muscle and heart express not only isoform 1, but also muscle-specific isoform mRNAs (isoforms 2, 3 and 4). Isoforms 5 and 6 are present in both liver and muscle

## **AGL Antibody (Center) - Protocols**

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

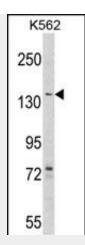
- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- <u>Immunoprecipitation</u>
- Flow Cytomety
- Cell Culture

# **AGL Antibody (Center) - Images**



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





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

# AGL Antibody (Center) - 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.

# **AGL Antibody (Center) - 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 (Center) - Citations**

- Loss of glycogen debranching enzyme AGL drives bladder tumor growth via induction of hyaluronic acid synthesis.
- Muscle glycogen remodeling and glycogen phosphate metabolism following exhaustive exercise of wild type and laforin knockout mice.
- Genetic depletion of the malin E3 ubiquitin ligase in mice leads to lafora bodies and the accumulation of insoluble laforin.
- 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.