

## GCS1 Antibody (C-term)

Purified Rabbit Polyclonal Antibody (Pab) Catalog # AP2315b

## **Specification**

## GCS1 Antibody (C-term) - Product Information

WB,E Application **Primary Accession** 013724 Other Accession NP 006293 Human, Mouse Reactivity Host **Rabbit** Clonality **Polyclonal** Isotype Rabbit IgG Calculated MW 91918 Antigen Region 796-826

## GCS1 Antibody (C-term) - Additional Information

#### **Gene ID 7841**

#### **Other Names**

Mannosyl-oligosaccharide glucosidase, Processing A-glucosidase I, MOGS, GCS1

#### Target/Specificity

This GCS1 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 796-826 amino acids from the C-terminal region of human GCS1.

#### **Dilution**

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**

GCS1 Antibody (C-term) is for research use only and not for use in diagnostic or therapeutic procedures.

## GCS1 Antibody (C-term) - Protein Information

## Name MOGS (HGNC:24862)

Function In the context of N-glycan degradation, cleaves the distal alpha 1,2-linked glucose



residue from the Glc(3)Man(9)GlcNAc(2) oligosaccharide precursor in a highly specific manner.

#### **Cellular Location**

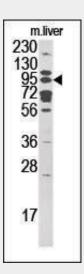
Endoplasmic reticulum membrane; Single-pass type II membrane protein {ECO:0000250|UniProtKB:088941}

## GCS1 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

## GCS1 Antibody (C-term) - Images



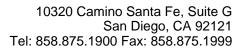
Western blot analysis of anti-GCS1 Pab (Cat. #AP2315b) in mouse liver cell line lysate (35ug/lane). GCS1(arrow) was detected using the purified Pab.

## GCS1 Antibody (C-term) - Background

GCS1 cleaves the distal alpha 1,2-linked glucose residue from the Glc(3)Man(9)GlcNAc(2) oligosaccharide precursor in a highly specific manner. Defects in GCS1 are the cause of type IIb congenital disorder of glycosylation (CDGIIb). This syndrome is also known as glucosidase I deficiency and is characterized by marked generalized hypotonia and hypomotility of the neonate, dysmorphic features, including a prominent occiput, short palpebral fissures, retrognathia, high arched palate, generalized edema, and hypoplastic genitalia. Symptoms include hepatomegaly, hypoventilation, feeding problems and seizures. The clinical course is progressive and survival is at most a few months.

# GCS1 Antibody (C-term) - References

Volker, C., et al., Glycobiology 12(8):473-483 (2002).





De Praeter, C.M., et al., Am. J. Hum. Genet. 66(6):1744-1756 (2000). Kalz-Fuller, B., et al., Eur. J. Biochem. 231(2):344-351 (1995).