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

GCS1 Antibody (N-term)
Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP2315a

Specification

GCS1 Antibody (N-term) - Product Information

Application	**WB,E**
Primary Accession	Q13724
Other Accession	NP_006293
Reactivity	**Human**
Host	**Rabbit**
Clonality	**Polyclonal**
Isotype	**Rabbit IgG**
Calculated MW	**91918**
Antigen Region	**1-30**

GCS1 Antibody (N-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 1-30 amino acids from the N-terminal region of human GCS1.

Dilution
WB~~1:1000

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

GCS1 Antibody (N-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.

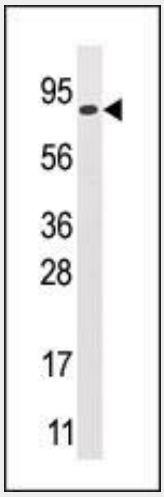
Cellular Location
Endoplasmic reticulum membrane; Single-pass type II membrane protein {ECO:0000250|UniProtKB:O88941}

GCS1 Antibody (N-term) - Protocols

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

- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

GCS1 Antibody (N-term) - Images



Western blot analysis of anti-GCS1 Pab (Cat. #AP2315a) in 293 cell line lysates (35ug/lane). GCS1 (arrow) was detected using the purified Pab.

GCS1 Antibody (N-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 (N-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).

Kalz-Fueller, B., et al., Eur. J. Biochem. 249, 912-912 (1997).