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GCS1 Antibody (C-term) Blocking Peptide
Synthetic peptide
Catalog # BP2315b

Specification

GCS1 Antibody (C-term) Blocking Peptide - Product Information

Primary Accession	Q13724
Other Accession	NP_006293

GCS1 Antibody (C-term) Blocking Peptide - Additional Information

Gene ID 7841

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

Target/Specificity
The synthetic peptide sequence used to generate the antibody AP2315b was selected from the C-term region of human GCS1 . A 10 to 100 fold molar excess to antibody is recommended. Precise conditions should be optimized for a particular assay.

Format
Peptides are lyophilized in a solid powder format. Peptides can be reconstituted in solution using the appropriate buffer as needed.

Storage
Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C.

Precautions
This product is for research use only. Not for use in diagnostic or therapeutic procedures.

GCS1 Antibody (C-term) Blocking Peptide - 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 (C-term) Blocking Peptide - Protocols

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

- Blocking Peptides

GCS1 Antibody (C-term) Blocking Peptide - Images

GCS1 Antibody (C-term) Blocking Peptide - 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) Blocking Peptide - 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).