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GCS1 Antibody (C-term)

Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP2315b

Product Information

Application	WB, E
Primary Accession	Q13724
Other Accession	NP_006293
Reactivity	Human, Mouse
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB4963/4964
Calculated MW	91918
Antigen Region	796-826

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.

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}

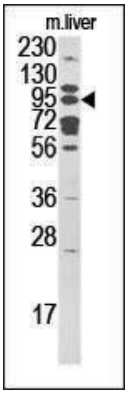
Background

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.

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).

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.