abcam

Product datasheet

Anti-GCS1 antibody ab82962

1 Image

Overview

Product name	Anti-GCS1 antibody
Description	Rabbit polyclonal to GCS1
Tested applications	**Suitable for:** WB, ELISA
Species reactivity	**Reacts with:** Human **Predicted to work with:** Mouse, Rat, Rabbit, Horse, Guinea pig, Cow, Cat, Dog, Zebrafish
Immunogen	Synthetic peptide corresponding to a region within N terminal amino acids 144-193 (GPYGWEFHDG LSFGRQHIQD GALRLTTEFV KRPGGQHGGD WSWRVTVEPQ) of human GCS1 (NP_006293). Run BLAST with ExPASy Run BLAST with NCBI
Positive control	HeLa cell lysate.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid repeated freeze / thaw cycles.
Storage buffer	Preservative: None Constituents: 2% Sucrose, PBS
Purity	Immunogen affinity purified
Purification notes	ab82962 is purified by a peptide affinity chromatography method.
Clonality	Polyclonal
Isotype	IgG

Applications

Our Abpromise guarantee covers the use of **ab82962** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes

Application	Abreviews	Notes
WB		Use a concentration of 1 µg/ml. Predicted molecular weight: 92 kDa. Good results were obtained when blocked with 5% non-fat dry milk in 0.05% PBS-T.
ELISA		Use at an assay dependent concentration. ELISA titre using peptide based assay, 1:312500.

Target

Function Cleaves the distal alpha 1,2-linked glucose residue from the Glc(3)Man(9)GlcNAc(2) oligosaccharide precursor in a highly specific manner.

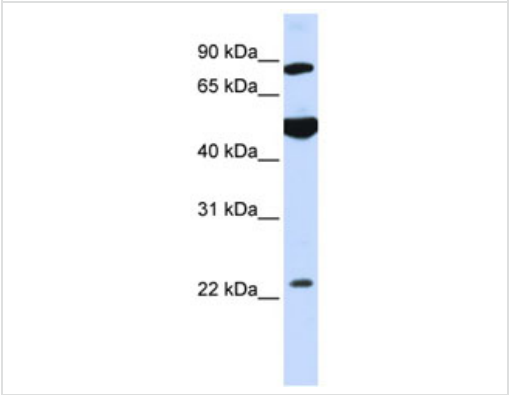
Pathway Glycan metabolism; N-glycan degradation.

Involvement in disease Defects in MOGS are the cause of type IIb congenital disorder of glycosylation (CDGIIb) [MIM:606056]; also known as glucosidase I deficiency. CDGIIb 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 of the infant included hepatomegaly, hypoventilation, feeding problems and seizures. The clinical course was progressive and the infant did not survive more than a few months.

Sequence similarities Belongs to the glycosyl hydrolase 63 family.

Cellular localization Endoplasmic reticulum membrane.

Images



Western blot - GCS1 antibody (ab82962)

Anti-GCS1 antibody (ab82962) at 1 µg/ml + HeLa cell lysate at 10 µg

Secondary
HRP conjugated anti-Rabbit IgG at 1/50000 dilution

Predicted band size : 92 kDa
Observed band size : 92 kDa
Additional bands at : 24 kDa,50 kDa. We are unsure as to the identity of these extra bands.

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