abcam

Product datasheet

Anti-GCS1 antibody ab82962

1 Image

Overview

Product name Anti-GCS1 antibody

Description Rabbit polyclonal to GCS1

Host species Rabbit

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

A

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 S 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.

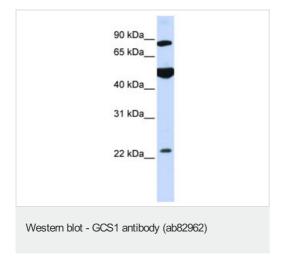
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Application	Abreviews	Notes
WB		Use a concentration of 1 μ g/ml. Predicted molecular weight: 92 kDa. Good results were obtained when blocked with 5% non-fat dry milk in 0.05% PBS-T.
ELISA		Use at an assay dependent concentration. ELISA titre using 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 Ilb congenital disorder of glycosylation (CDGIlb) [MIM:606056]; also known as glucosidase I deficiency. CDGIlb 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



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

Secondary

HRP conjugated anti-Rabbit lgG 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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