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Product datasheet for AP12152PU-N

GCS1 (MOGS) (N-term) Rabbit Polyclonal Antibody

Product data:

Product Type:	Primary Antibodies
Applications:	WB
Recommended Dilution:	ELISA: 1/1,000. Western Blot: 1/50-1/100.
Reactivity:	Human
Host:	Rabbit
Isotype:	Ig
Clonality:	Polyclonal
Immunogen:	This antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide selected from the N-terminal region of human GCS1.
Specificity:	This antibody is specific to GCS1 (N-term).
Formulation:	PBS with 0.09% (W/V) Sodium Azide as preservative. State: Purified State: Liquid purified Ig fraction.
Concentration:	lot specific
Purification:	Protein G Chromatography, eluted with high and low pH buffers and neutralized immediately, followed by dialysis against PBS.
Conjugation:	Unconjugated
Storage:	Store the antibody undiluted at 2-8°C for one month or (in aliquots) at -20°C for longer. Avoid repeated freezing and thawing.
Stability:	Shelf life: one year from despatch.
Gene Name:	mannosyl-oligosaccharide glucosidase
Database Link:	Entrez Gene 7841 Human Q13724

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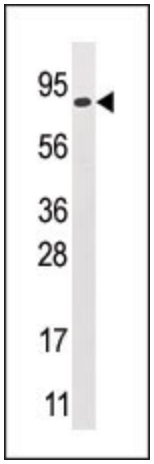
Background: GCS1 cleaves the distal alpha 1,2-linked glucose residue from the Glc(3)Man(9)GlcNAc(2) oligosaccharide precursor in a highly specific manner. Defects in GCS1 are the cause of type IIb congenital disorder of glycosylation (CDGIIb). This syndrome is also known as glucosidase I deficiency and 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.

Synonyms: GCS1

Note: **Predicted Molecular weight:** 91840 Da

Product images:



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