

PMM2 Antibody (monoclonal) (M01)

Mouse monoclonal antibody raised against a partial recombinant PMM2. Catalog # AT3355a

Product Information

Application WB, E **Primary Accession** 015305 **Other Accession** NM 000303 Reactivity Human Host mouse Clonality monoclonal Isotype IgG2b Kappa **Clone Names** 2E10 Calculated MW 28082

Additional Information

Gene ID 5373

Other Names Phosphomannomutase 2, PMM 2, PMM2

Target/Specificity PMM2 (NP_000294, 47 a.a. ~ 111 a.a) partial recombinant protein with GST

tag. MW of the GST tag alone is 26 KDa.

Dilution WB~~1:500~1000 E~~N/A

Format Clear, colorless solution in phosphate buffered saline, pH 7.2.

Storage Store at -20°C or lower. Aliquot to avoid repeated freezing and thawing.

Precautions PMM2 Antibody (monoclonal) (M01) is for research use only and not for use

in diagnostic or therapeutic procedures.

Background

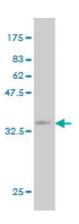
The protein encoded by this gene catalyzes the isomerization of mannose 6-phosphate to mannose 1-phosphate, which is a precursor to GDP-mannose necessary for the synthesis of dolichol-P-oligosaccharides. Mutations in this gene have been shown to cause defects in glycoprotein biosynthesis, which manifests as carbohydrate-deficient glycoprotein syndrome type I.

References

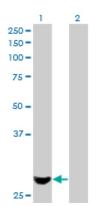
Functional analysis of three splicing mutations identified in the PMM2 gene: toward a new therapy for congenital disorder of glycosylation type Ia. Vega AI, et al. Hum Mutat, 2009 May. PMID 19235233.Primary skeletal dysplasia as a major manifesting feature in an infant with congenital disorder of glycosylation type

Ia. Coman D, et al. Am J Med Genet A, 2008 Feb 1. PMID 18203160. Cerebellar ataxia and congenital disorder of glycosylation Ia (CDG-Ia) with normal routine CDG screening. Vermeer S, et al. J Neurol, 2007 Oct. PMID 17694350. Congenital disorder of glycosylation type 1a: three siblings with a mild neurological phenotype. Coman D, et al. J Clin Neurosci, 2007 Jul. PMID 17451957. Characterization of two unusual truncating PMM2 mutations in two CDG-Ia patients. Schollen E, et al. Mol Genet Metab, 2007 Apr. PMID 17307006.

Images

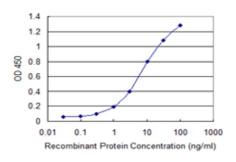


Antibody Reactive Against Recombinant Protein.Western Blot detection against Immunogen (32.89 KDa) .



Western Blot analysis of PMM2 expression in transfected 293T cell line by PMM2 monoclonal antibody (M01), clone 2E9.

Lane 1: PMM2 transfected lysate(28.1 KDa). Lane 2: Non-transfected lysate.



Detection limit for recombinant GST tagged PMM2 is 0.1 ng/ml as a capture antibody.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.