



PMGT1 rabbit pAb

Catalog No	BYab-10946
Isotype	IgG
Reactivity	Human; Mouse;Rat
Applications	WB;IHC
Gene Name	POMGNT1 MGAT1.2 UNQ746/PRO1475
Protein Name	PMGT1
Immunogen	Synthesized peptide derived from human PMGT1 AA range: 171-221
Specificity	This antibody detects endogenous levels of PMGT1 at Human/Mouse/Rat
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit serum by affinity-chromatography using specific immunogen.
Dilution	WB 1:500-2000;IHC-p 1:50-300
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	
Observed Band	
Cell Pathway	Golgi apparatus membrane ; Single-pass type II membrane protein .
Tissue Specificity	Constitutively expressed. An additional weaker band is also detected in spinal cord, lymph node, and trachea. Expressed especially in astrocytes. Also expressed in immature and mature neurons.
Function	catalytic activity:UDP-N-acetyl-D-glucosamine + Man-R = N-acetyl-D-glucosamine-beta-1,2-Man-R + UDP.,cofactor:Manganese.,disease:Defects in POMGNT1 are a cause of Walker-Warburg syndrome (WWS) [MIM:236670]; also known as hydrocephalus-agyria-retinal dysplasia or HARD syndrome. WWS is an autosomal recessive disorder characterized by cobblestone lissencephaly, hydrocephalus, agyria, retinal dysplasia, with or without encephalocele. It is often associated with congenital muscular dystrophy and usually lethal within the first few months of life.,disease:Defects in POMGNT1 are the cause of muscle-eye-brain disease (MEB) [MIM:253280]. MEB is an autosomal recessive disorder characterized by congenital muscular dystrophy, ocular abnormalities,

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cobblestone lissencephaly and cerebellar hypoplasia. MEB patients present severe congenital myopia, congenital glaucoma, pallor of the optic disks, retina

Background

This gene encodes a type II transmembrane protein that resides in the Golgi apparatus. It participates in O-mannosyl glycosylation and is specific for alpha linked terminal mannose. Mutations in this gene may be associated with muscle-eye-brain disease and several congenital muscular dystrophies. Alternatively spliced transcript variants that encode different protein isoforms have been described. [provided by RefSeq, Feb 2014],

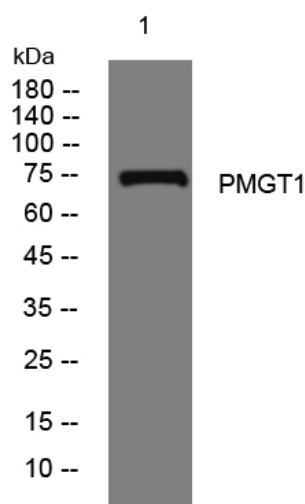
matters needing attention

Avoid repeated freezing and thawing!

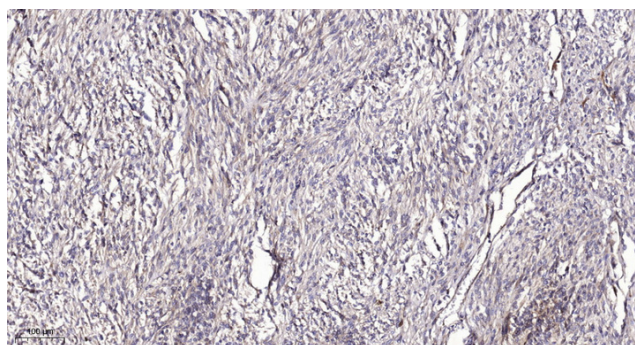
Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western blot analysis of lysates from 293T cells, primary antibody was diluted at 1:1000, 4° over night



Immunohistochemical analysis of paraffin-embedded human Colon cancer. 1, Antibody was diluted at 1:200(4° overnight). 2, Tris-EDTA, pH9.0 was used for antigen retrieval. 3, Secondary antibody was diluted at 1:200(room temperature, 45min).