10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

Fukutin Recombinant Rabbit mAb

Catalog # AP100046

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	O75072
Other Accession	O75072
Reactivity	Rat, Human, Mouse
Host	Rabbit
Clonality	Recombinant
Calculated MW	53724
Physical State	Liquid
Immunogen	A synthesized peptide derived from human Fukutin
Epitope Specificity	350-461
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	10mM phosphate buffered saline(pH 7.4) with 150mM sodium chloride, 0.05% BSA, 0.02% Proclin300 and 50% glycerol.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Catalyzes the transfer of a ribitol-phosphate from CDP-ribitol to the distal N-acetylgalactosamine of the phosphorylated O-mannosyl trisaccharide (N-acetylgalactosamine-beta-3-N-acetylglucosamine-beta-4-(phosphate-6-)mannose), a carbohydrate structure present in alpha-dystroglycan (DAG1).

Additional Information

Gene ID	2218
Other Names	FKTN_HUMAN; FKTN; Fukutin; Fukuyama-type congenital muscular dystrophy protein; Ribitol-5-phosphate transferase; 2.7.8.-; FCMD; CMD1X; LGMD2M; LGMDR13; MDDGA4; MDDGB4; MDDGC4; Fukuyama type congenital muscular dystrophy (fukutin)
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	FKTN (HGNC:3622)
Function	Catalyzes the transfer of a ribitol-phosphate from CDP- ribitol to the distal

	N-acetylgalactosamine of the phosphorylated O- mannosyl trisaccharide (N-acetylgalactosamine-beta-3-N-acetylglucosamine-beta-4-(phosphate-6-)mannose), a carbohydrate structure present in alpha-dystroglycan (DAG1) (PubMed:26923585, PubMed:27194101, PubMed:29477842). This constitutes the first step in the formation of the ribitol 5-phosphate tandem repeat which links the phosphorylated O-mannosyl trisaccharide to the ligand binding moiety composed of repeats of 3-xylosyl-alpha-1,3-glucuronic acid-beta-1 (PubMed:17034757, PubMed:25279699, PubMed:26923585, PubMed:27194101, PubMed:29477842). Required for normal location of POMGNT1 in Golgi membranes, and for normal POMGNT1 activity (PubMed:17034757). May interact with and reinforce a large complex encompassing the outside and inside of muscle membranes (PubMed:25279699). Could be involved in brain development (Probable).
Cellular Location	Golgi apparatus membrane; Single-pass type II membrane protein. Cytoplasm {ECO:0000250|UniProtKB:Q8R507}. Nucleus {ECO:0000250|UniProtKB:Q8R507}. Note=In retinal tissue, does not localize with the Golgi apparatus. {ECO:0000250|UniProtKB:Q8R507}
Tissue Location	Expressed in the retina (at protein level) (PubMed:29416295). Widely expressed with highest expression in brain, heart, pancreas and skeletal muscle (PubMed:11115853). Expressed at similar levels in control fetal and adult brain (PubMed:11115853) Expressed in migrating neurons, including Cajar-Retzius cells and adult cortical neurons, as well as hippocampal pyramidal cells and cerebellar Purkinje cells (PubMed:11115853). No expression observed in the glia limitans, the subpial astrocytes (which contribute to basement membrane formation) or other glial cells (PubMed:11115853)

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