

FKRP Antibody (Center) Blocking Peptide
Synthetic peptide
Catalog # BP14285c**Specification**

FKRP Antibody (Center) Blocking Peptide - Product InformationPrimary Accession [Q9H9S5](#)**FKRP Antibody (Center) Blocking Peptide - Additional Information**

Gene ID 79147

Other Names

Fukutin-related protein, 2---, FKRP

Format

Peptides are lyophilized in a solid powder format. Peptides can be reconstituted in solution using the appropriate buffer as needed.

Storage

Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C.

Precautions

This product is for research use only. Not for use in diagnostic or therapeutic procedures.

FKRP Antibody (Center) Blocking Peptide - Protein InformationName FKRP ([HGNC:17997](#))**Function**

Catalyzes the transfer of a ribitol 5-phosphate from CDP-L- ribitol to the ribitol 5-phosphate previously attached by FKTN/fukutin to 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:29477842, PubMed:31949166, PubMed:27194101). This constitutes the second step in the formation of the ribose 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:25279699, PubMed:26923585, PubMed:29477842, PubMed:31949166, PubMed:27194101).

Cellular Location

Golgi apparatus membrane; Single-pass type II membrane protein. Secreted. Cell membrane, sarcolemma {ECO:0000250|UniProtKB:Q8CG64}. Rough endoplasmic reticulum. Cytoplasm {ECO:0000250|UniProtKB:Q8CG64}. Note=According to some studies the N- terminal hydrophobic domain is cleaved after translocation to the Golgi apparatus and the protein is secreted (PubMed:19900540). Localization at the cell membrane may require the presence of dystroglycan (By similarity). At the Golgi apparatus localizes to the middle-to-trans- cisternae, as assessed by MG160 colocalization. Detected in rough endoplasmic reticulum in myocytes (PubMed:17554798, PubMed:21886772) In general, mutants associated with severe clinical phenotypes are retained within the endoplasmic reticulum (PubMed:15213246) {ECO:0000250|UniProtKB:Q8CG64, ECO:0000269|PubMed:15213246, ECO:0000269|PubMed:17554798, ECO:0000269|PubMed:19900540, ECO:0000269|PubMed:21886772}

Tissue Location

Expressed in the retina (at protein level) (PubMed:29416295). Expressed predominantly in skeletal muscle, placenta, and heart and relatively weakly in brain, lung, liver, kidney, and pancreas (PubMed:11592034).

FKRP Antibody (Center) Blocking Peptide - Protocols

Provided below are standard protocols that you may find useful for product applications.

- [Blocking Peptides](#)

FKRP Antibody (Center) Blocking Peptide - Images

FKRP Antibody (Center) Blocking Peptide - Background

This gene encodes a protein which is targeted to the medial Golgi apparatus and is necessary for posttranslational modification of dystroglycan. Mutations in this gene have been associated with congenital muscular dystrophy, mental retardation, and cerebellar cysts. Several alternatively spliced transcript variants of this gene have been described, but the full-length nature of some of these variants has not been determined. [provided by RefSeq].

FKRP Antibody (Center) Blocking Peptide - References

Kawahara, G., et al. Hum. Mol. Genet. 19(4):623-633(2010) Crowther-Swanepoel, D., et al. Nat. Genet. 42(2):132-136(2010) Lu, P.J., et al. Biochim. Biophys. Acta 1802(2):253-258(2010) Hanisch, F., et al. J. Neurol. 257(2):300-301(2010) Bourteel, H., et al. J. Neurol. Neurosurg. Psychiatr. 80(12):1405-1408(2009)