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FKRP Antibody (C-term)
Mouse Monoclonal Antibody (Mab)
Catalog # AM2033b

Specification

FKRP Antibody (C-term) - Product Information

Application	**WB,E**
Primary Accession	Q9H9S5
Other Accession	Q8CG64, NP_001034974.1
Reactivity	**Human**
Predicted	**Mouse**
Host	**Mouse**
Clonality	**Monoclonal**
Isotype	**IgG1**
Calculated MW	**54568**
Antigen Region	**418-444**

FKRP Antibody (C-term) - Additional Information

Gene ID 79147

Other Names
Fukutin-related protein, 2---, FKRP

Target/Specificity
This FKRP antibody is generated from mice immunized with a KLH conjugated synthetic peptide between 418-444 amino acids from the C-terminal region of human FKRP.

Dilution
WB~~1:100

Format
Purified monoclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein G column, followed by dialysis against PBS.

Storage
Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions
FKRP Antibody (C-term) is for research use only and not for use in diagnostic or therapeutic procedures.

FKRP Antibody (C-term) - Protein Information

Name 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:27194101, PubMed:29477842, PubMed:31949166). 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:27194101, PubMed:29477842, PubMed:31949166).

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 (C-term) - Protocols

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

- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

FKRP Antibody (C-term) - Images

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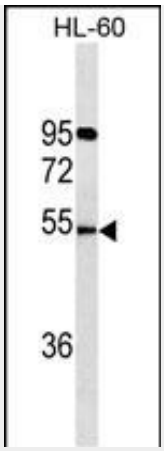

FKRP Antibody (C-term) (Cat. #AM2033b) western blot analysis in HL-60 cell line lysates (35µg/lane).This demonstrates the FKRP antibody detected the FKRP protein (arrow).

FKRP Antibody (C-term) - 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 (C-term) - 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)