

FKRP (E-4): sc-374642

BACKGROUND

Fukutin-related protein (FKRP) is ubiquitously expressed, with highest expression in heart, skeletal muscle and placenta, and weakest expression in lung, liver, brain, kidney and pancreas. FKRP localizes to the medial Golgi apparatus through its N-terminal and transmembrane domains. It is a predicted glycosyltransferase protein that plays a role in α -dystroglycan glycosylation. Mutations in FKRP cause various diseases including congenital muscular dystrophy 1C (MDC1C), limb-girdle muscular dystrophy type 2I (LGMD2I) and congenital muscular dystrophies (CMDs) with brain malformations and mental retardation. FKRP mutations may also cause muscle-eye-brain disease (MEB) and Walker-Warburg syndrome (WWS), disorders characterized by disruption of brain and eye structure in addition to muscular dystrophy. Mislocalization of FKRP from the Golgi apparatus is a potential result of mutations in FKRP.

REFERENCES

1. Brockington, M., et al. 2001. Mutations in the Fukutin-related protein gene muscular dystrophy with secondary laminin α 2 deficiency and abnormal glycosylation of α -dystroglycan. *Am. J. Hum. Genet.* 69: 1198-1209.
2. Online Mendelian Inheritance in Man, OMIM[™]. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 606596. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
3. Esapa, C.T., et al. 2004. Fukutin-related protein mutations that cause congenital muscular dystrophy result in ER-retention of the mutant protein in cultured cells. *Hum. Mol. Genet.* 14: 295-305.
4. Müller, T., et al. 2005. Dilated cardiomyopathy may be an early sign of the C826A Fukutin-related protein mutation. *Neuromuscul. Disord.* 15: 372-376.
5. Dolatshad, N.F., et al. 2005. Mutated Fukutin-related protein (FKRP) localises as wildtype in differentiated muscle cells. *Exp. Cell Res.* 309: 370-378.

CHROMOSOMAL LOCATION

Genetic locus: FKRP (human) mapping to 19q13.32; Fkrp (mouse) mapping to 7 A2.

SOURCE

FKRP (E-4) is a mouse monoclonal antibody raised against amino acids 368-495 mapping at the C-terminus of FKRP of human origin.

PRODUCT

Each vial contains 200 μ g IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

FKRP (E-4) is available conjugated to agarose (sc-374642 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-374642 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-374642 PE), fluorescein (sc-374642 FITC), Alexa Fluor[®] 488 (sc-374642 AF488), Alexa Fluor[®] 546 (sc-374642 AF546), Alexa Fluor[®] 594 (sc-374642 AF594) or Alexa Fluor[®] 647 (sc-374642 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor[®] 680 (sc-374642 AF680) or Alexa Fluor[®] 790 (sc-374642 AF790), 200 μ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

FKRP (E-4) is recommended for detection of FKRP of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for FKRP siRNA (h): sc-60645, FKRP siRNA (m): sc-60646, FKRP shRNA Plasmid (h): sc-60645-SH, FKRP shRNA Plasmid (m): sc-60646-SH, FKRP shRNA (h) Lentiviral Particles: sc-60645-V and FKRP shRNA (m) Lentiviral Particles: sc-60646-V.

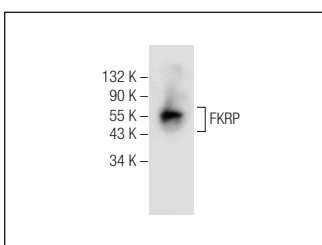
Molecular Weight of FKRP: 60 kDa.

Positive Controls: human skeletal muscle extract: sc-363776.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker[™] Molecular Weight Standards: sc-2035, UltraCruz[®] Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz[®] Mounting Medium: sc-24941 or UltraCruz[®] Hard-set Mounting Medium: sc-359850.

DATA



FKRP (E-4): sc-374642. Western blot analysis of FKRP expression in human skeletal muscle tissue extract.

SELECT PRODUCT CITATIONS

1. Badolia, R., et al. 2020. The role of non-glycolytic glucose metabolism in myocardial recovery upon mechanical unloading and circulatory support in chronic heart failure. *Circulation* 142: 259-274.

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

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