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 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**


**CHROMOSOMAL LOCATION**

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

**SOURCE**

FKRP (E-4) is a mouse monoclonal antibody raised against amino acids 388-495 mapping at the C-terminus of FKRP of human origin.

**PRODUCT**

Each vial contains 200 µg 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 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-374642 HRP), 200 µg/ml, for WB, IHC(+) 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 µg/ml, for WB (RGB), IF, IHC(+) and FCM; and to either Alexa Fluor® 680 (sc-374642 AF680) or Alexa Fluor® 790 (sc-374642 AF790), 200 µg/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 µg per 100-500 µg 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-60646-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 BP-HRP: sc-516102 or m-IgG BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz MarkerTM 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 BP-FITC: sc-516214 or m-IgG BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Hard-set Mounting Medium: sc-24941 or UltraCruz® 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**


**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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