SANTA CRUZ BIOTECHNOLOGY, INC.

FKRP (H-128): sc-292627

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The Power to Question

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 α2 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.
6. Boito, C.A., et al. 2005. Clinical and molecular characterization of patients with limb-girdle muscular dystrophy type 2I. Arch. Neurol. 62: 1894-1899.
7. Vajsar, J. and Schachter, H. 2006. Walker-Warburg syndrome. Orphanet J. Rare Dis. 1: 29.

CHROMOSOMAL LOCATION

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

SOURCE

FKRP (H-128) is a rabbit polyclonal antibody raised against amino acids 368-495 mapping at the C-terminus of FKRP of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

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

APPLICATIONS

FKRP (H-128) is recommended for detection of FKRP of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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).

FKRP (H-128) is also recommended for detection of FKRP in additional species, including canine, bovine and porcine.

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

Molecular Weight of FKRP: 60 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.