SANTA CRUZ BIOTECHNOLOGY, INC.

FKRP (K-20): sc-48505

SANTA CRUZ BIOTECHNOLOGY

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 (K-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-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.

Blocking peptide available for competition studies, sc-48505 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

FKRP (K-20) 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 (K-20) is also recommended for detection of FKRP in additional species, including canine, bovine and porcine.

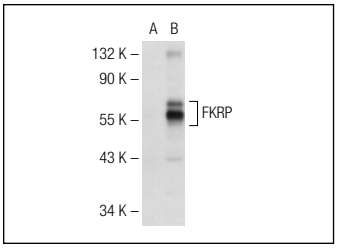
Suitable for use as control antibody for FKRP siRNA (h): sc-60645, FKRP siRNA (m): sc-60646, FKRP shRNA Plasmid (h): sc-60645-SH, FKRP shRNA Plasmid (m): sc-60646-SH, FKRP shRNA (h) Lentiviral Particles: sc-60645-V and FKRP shRNA (m) Lentiviral Particles: sc-60646-V.

FKRP (K-20) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

Molecular Weight of FKRP: 60 kDa.

Positive Controls: FKRP (h): 293 Lysate: sc-159806.

DATA



FKRP (K-20): sc-48505. Western blot analysis of FKRP expression in non-transfected: sc-110760 (**A**) and human FKRP transfected: sc-159806 (**B**) 293 whole cell lysates.

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.

PROTOCOLS

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