SANTA CRUZ BIOTECHNOLOGY, INC.

Fukutin (H-300): sc-134573



BACKGROUND

Fukutin, a secreted protein, is expressed in various tissues in normal individuals. Fukutin co-localizes with a Golgi marker and a granular cytoplasmic distribution, suggesting that it passes through the Golgi before being packaged into secretory vesicles. Fukutin may be located in the extracellular matrix, where it interacts with and reinforces a large complex encompassing the outside and inside of muscle membranes; alternatively, as a secreted protein, Fukutin may cause muscular dystrophy by an unknown mechanism. The Fukutin gene is expressed at similar levels in control fetal and adult brain, but is much reduced in Fukuyama congenital muscular dystrophy (FCMD) brain. Fukutin deficiency affects the modification of glycosylation of DAG1 (α -dystroglycan), which then cannot localize or function properly and may be degraded or eluted from the extracellular surface membrane of the muscle fiber. FCMD is the first human disease known to be caused by an ancient retrotransposal integration. The gene which encodes Fukutin maps to human chromosome 9q31.2.

REFERENCES

- Toda, T., et al. 1996. Linkage-disequilibrium mapping narrows the Fukuyamatype congenital muscular dystrophy (FCMD) candidate region to less than 100 kb. Am. J. Hum. Genet. 59: 1313-1320.
- 2. Kobayashi, K., et al. 1998. An ancient retrotransposal insertion causes Fukuyama-type congenital muscular dystrophy. Nature 394: 388-392.
- Sasaki, J., et al. 2000. Neuronal expression of the Fukutin gene. Hum. Mol. Genet. 9: 3083-3090.
- Hayashi, Y.K., et al. 2001. Selective deficiency of α-dystroglycan in Fukuyama-type congenital muscular dystrophy. Neurology 57: 115-121.
- 5. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 607440. World Wide Web URL: http://www.ncbi.nlm. nih.gov/omim/
- Chiyonobu, T., et al. 2005. Effects of Fukutin deficiency in the developing mouse brain. Neuromuscul. Disord. 15: 416-426.
- 7. Fang, H., et al. 2005. Identification of a functional CRE in the promoter of Fukuyama congenital muscular dystrophy gene Fukutin. Brain Res. Mol. Brain Res. 136: 1-11.
- Kurahashi, H., et al. 2005. Basement membrane fragility underlies embryonic lethality in Fukutin-null mice. Neurobiol. Dis. 19: 208-217.

CHROMOSOMAL LOCATION

Genetic locus: FKTN (human) mapping to 9q31.2; Fktn (mouse) mapping to 4 B2.

SOURCE

Fukutin (H-300) is a rabbit polyclonal antibody raised against amino acids 1-300 mapping at the N-terminus of Fukutin of human origin.

PRODUCT

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

APPLICATIONS

Fukutin (H-300) is recommended for detection of Fukutin of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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).

Fukutin (H-300) is also recommended for detection of Fukutin in additional species, including equine, canine and bovine.

Suitable for use as control antibody for Fukutin siRNA (h): sc-43773, Fukutin siRNA (m): sc-60664, Fukutin shRNA Plasmid (h): sc-43773-SH, Fukutin shRNA Plasmid (m): sc-60664-SH, Fukutin shRNA (h) Lentiviral Particles: sc-43773-V and Fukutin shRNA (m) Lentiviral Particles: sc-60664-V.

Molecular Weight of Fukutin: 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.

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.