SANTA CRUZ BIOTECHNOLOGY, INC.

FRG1 (L-07): sc-101050

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

FRG1 is a 258 amino acid nuclear protein encoded by the human gene FRG1. The FRG1 protein is thought to be involved in pre-messenger RNA splicing. FRG1 plays a role in processing pre-rRNA, assembling rRNA into ribosomal subunits and may also be involved in pre-mRNA splicing. Facioscapulohumeral muscular dystrophy (FSHD) is a disease state associated with internal deletions among the tandem array of D4Z4 repeats on chromosome 4q35, a subtelomere region of chromosome 4 that contains the FRG1 gene. The muscle degeneration that is common in patients with FSHD results from increased expression of genes proximal to the deletion, including FRG1. In addition to muscle degeneration, most FSHD patients also develop abnormalities of the retinal vasculature. FRG1 is expressed in adult and fetal muscle, lymphocytes and placenta. It can be localized to nuclear Cajal bodies or speckles.

REFERENCES

1. van Deutekom, J.C., et al. 1996. Identification of the first gene (FRG1) from the FSHD region on human chromosome 4q35. Hum. Mol. Genet. 5: 581-590.
2. Grewal, P.K., et al. 1997. The mouse homolog of FRG1, a candidate gene for FSHD, maps proximal to the myodystrophy mutation on chromosome 8. Mamm. Genome 8: 394-398.

CHROMOSOMAL LOCATION

Genetic locus: FRG1 (human) mapping to 4q35.2; Frg1 (mouse) mapping to 8 A4.

SOURCE

FRG1 (L-07) is a mouse monoclonal antibody raised against recombinant FRG1 of human origin.

PRODUCT

Each vial contains 100 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

FRG1 (L-07) is recommended for detection of FRG1 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).

Suitable for use as control antibody for FRG1 siRNA (h): sc-62350, FRG1 siRNA (m): sc-62351, FRG1 shRNA Plasmid (h): sc-62350-SH, FRG1 shRNA Plasmid (m): sc-62351-SH, FRG1 shRNA (h) Lentiviral Particles: sc-62350-V and FRG1 shRNA (m) Lentiviral Particles: sc-62351-V.

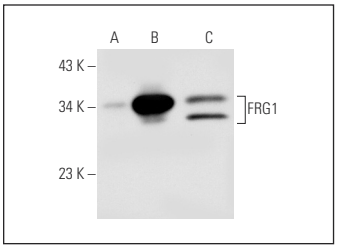
Molecular Weight of FRG1: 29 kDa.

Positive Controls: FRG1 (m): 293T Lysate: sc-120320, HeLa whole cell lysate: sc-2200 or FRG1 (h): 293T Lysate: sc-116506.

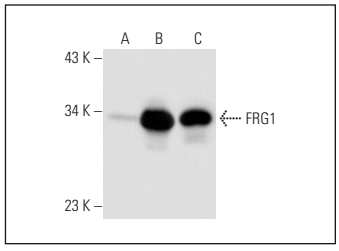
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ 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-IgGκ BP-FITC: sc-516140 or m-IgGκ 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



FRG1 (L-07): sc-101050. Western blot analysis of FRG1 expression in non-transfected 293T: sc-117752 (**A**), mouse FRG1 transfected 293T: sc-120320 (**B**) and HeLa (**C**) whole cell lysates.



FRG1 (L-07): sc-101050. Western blot analysis of FRG1 expression in non-transfected: sc-117752 (**A**) and human FRG1 transfected: sc-116506 (**B**) 293T whole cell lysates and HeLa nuclear extract (**C**).

SELECT PRODUCT CITATIONS

1. Bortolanza, S., et al. 2011. AAV6-mediated systemic shRNA delivery reverses disease in a mouse model of facioscapulohumeral muscular dystrophy. Mol. Ther. 19: 2055-2064.
2. Neguembor, M.V., et al. 2013. FSHD muscular dystrophy region gene 1 binds Suv4-20h1 histone methyltransferase and impairs myogenesis. J. Mol. Cell. Biol. 5: 294-307.
3. Pistoni, M., et al. 2013. Rbfox1 downregulation and altered calpain 3 splicing by FRG1 in a mouse model of facioscapulohumeral muscular dystrophy (FSHD). PLoS Genet. 9: e1003186.

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 for detailed protocols and support products.