

# FRG1 (N-19): sc-54594

## 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. 1997. 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.
3. Grewal, P.K., et al. 1998. FRG1, a gene in the FSH muscular dystrophy region on human chromosome 4q35, is highly conserved in vertebrates and invertebrates. *Gene* 216: 13-19.
4. Grewal, P.K., et al. 1999. Recent amplification of the human FRG1 gene during primate evolution. *Gene* 227: 79-88.
5. Tam, R., et al. 2004. The 4q subtelomere harboring the FSHD locus is specifically anchored with peripheral heterochromatin unlike most human telomeres. *J. Cell Biol.* 167: 269-279.
6. van Koningsbruggen, S., et al. 2004. FRG1P is localised in the nucleolus, Cajal bodies, and speckles. *J. Med. Genet.* 41: e46.
7. Gabellini, D., et al. 2006. Facioscapulohumeral muscular dystrophy in mice over-expressing FRG1. *Nature* 439: 973-977.
8. Osborne, R.J., et al. 2007. Expression profile of FSHD supports a link between retinal vasculopathy and muscular dystrophy. *Neurology* 68: 569-577.

## CHROMOSOMAL LOCATION

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

## SOURCE

FRG1 (N-19) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the N-terminus of FRG1 of human origin.

## RESEARCH USE

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

## 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-54594 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

FRG1 (N-19) 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).

FRG1 (N-19) is also recommended for detection of FRG1 in additional species, including porcine.

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: HeLa whole cell lysate: sc-2200 or HeLa nuclear extract: sc-2120.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (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 donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (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.

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.



Try **FRG1 (C-5): sc-377040** or **FRG1 (L-07): sc-101050**, our highly recommended monoclonal alternatives to FRG1 (N-19).