

PRPF3 (B-1): sc-518271



The Power to Question

BACKGROUND

PRPF3 (PRP3 pre-mRNA processing factor 3 homolog), also known as RP18, PRP3, Prp3p, HPRP3 or HPRP3P, is an evolutionarily conserved protein involved in pre-mRNA splicing and functions as a component of the U4/U6.U5 tri-snRNP (small nuclear ribonucleoprotein) complex. Ubiquitously expressed with predominant expression in retina, blood, kidney and liver, PRPF3 localizes to nuclear speckles and is phosphorylated *in vitro*. PRPF3 directly interacts with PRPF4 and is present in the inactive spliceosome but is not found in the catalytically active spliceosome. Mutations in the gene encoding PRPF3 result in autosomal dominant retinitis pigmentosa type 18 (RP18), which leads to photoreceptor cell degeneration. RP18 patients initially exhibit a loss of their midperipheral visual field as well as night vision blindness. The disease eventually progresses to the loss of far peripheral visual field and finally the loss of central vision. This suggests that PRPF3 is a key player in the pre-mRNA splicing of photoreceptor-specific genes.

REFERENCES

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- Chakarova, C.F., et al. 2002. Mutations in HPRP3, a third member of pre-mRNA splicing factor genes, implicated in autosomal dominant retinitis pigmentosa. *Hum. Mol. Genet.* 11: 87-92.
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- Comitato, A., et al. 2007. Mutations in splicing factor PRPF3, causing retinal degeneration, form detrimental aggregates in photoreceptor cells. *Hum. Mol. Genet.* 16: 1699-1707.
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CHROMOSOMAL LOCATION

Genetic locus: PRPF3 (human) mapping to 1q21.2.

SOURCE

PRPF3 (B-1) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 648-674 of PRPF3 of human origin.

PRODUCT

Each vial contains 200 µg IgM kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

PRPF3 (B-1) is recommended for detection of PRPF3 of human origin by Western Blotting (starting dilution 1:100, 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 PRPF3 siRNA (h): sc-88819, PRPF3 shRNA Plasmid (h): sc-88819-SH and PRPF3 shRNA (h) Lentiviral Particles: sc-88819-V.

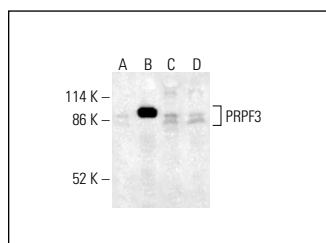
Molecular Weight of PRPF3: 77 kDa.

Positive Controls: PRPF3 (h): 293T Lysate: sc-173051 or Hep G2 cell lysate: sc-2227.

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 L-Agarose: sc-2336 (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



PRPF3 (B-1): sc-518271. Western blot analysis of PRPF3 expression in non-transfected 293T: sc-117752 (A), human PRPF3 transfected 293T: sc-173051 (B) and Hep G2 (D) whole cell lysates and HeLa nuclear extract (C). Detection reagent used: m-IgGκ BP-HRP: sc-516102.

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.