

PRPF31 (m2): 293T Lysate: sc-127396

BACKGROUND

PRPF31 (PRP31 pre-mRNA processing factor 31 homolog), also known as RP11, PRP31 or NY-BR-99, is a ubiquitously expressed protein that localizes to the nucleus and is found in Cajal bodies and speckles. PRPF31 is involved in pre-mRNA splicing and functions as a component of the U4/U6.U5 tri-snRNP (small nuclear ribonucleoprotein) complex. More specifically, PRPF31 is believed to mediate the tethering of the tri-snRNP to the spliceosome (a large ribonucleoprotein responsible for catalyzing the splicing of pre-mRNA), thereby assisting in the assembly of the mature spliceosome. Mutations in the gene encoding PRPF31 result in autosomal dominant retinitis pigmentosa type 11 (RP11), which leads to photoreceptor cell degeneration. RP11 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 PRPF31 is a key player in the pre-mRNA splicing of photoreceptor-specific genes.

REFERENCES

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2. Schaffert, N., et al. 2004. RNAi knockdown of hPrp 31 leads to an accumulation of U4/U6 di-snRNPs in Cajal bodies. *EMBO J.* 23: 3000-3009.
3. Yuan, L., et al. 2005. Mutations in PRPF31 inhibit pre-mRNA splicing of rhodopsin gene and cause apoptosis of retinal cells. *J. Neurosci.* 25: 748-757.
4. Gandra, M., et al. 2005. Gene symbol: PRPF31. Disease: retinitis pigmentosa—autosomal dominant. *Hum. Genet.* 118: 548-548.
5. Wilkie, S.E., et al. 2006. A study of the nuclear trafficking of the splicing factor protein PRPF31 linked to autosomal dominant retinitis pigmentosa (ADRP). *Biochim. Biophys. Acta* 1762: 304-311.
6. Rivolta, C., et al. 2006. Variation in retinitis pigmentosa-11 (PRPF31 or RP11) gene expression between symptomatic and asymptomatic patients with dominant RP11 mutations. *Hum. Mutat.* 27: 644-653.
7. Abu-Safieh, L., et al. 2006. A large deletion in the adRP gene PRPF31: evidence that haploinsufficiency is the cause of disease. *Mol. Vis.* 12: 384-388.
8. Mordes, D., et al. 2007. Identification of photoreceptor genes affected by PRPF31 mutations associated with autosomal dominant retinitis pigmentosa. *Neurobiol. Dis.* 26: 291-300.
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STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

PROTOCOLS

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

CHROMOSOMAL LOCATION

Genetic locus: Prpf31 (mouse) mapping to 7 A1.

PRODUCT

PRPF31 (m2): 293T Lysate represents a lysate of mouse PRPF31 transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

PRPF31 (m2): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive PRPF31 antibodies. Recommended use: 10-20 µl per lane.

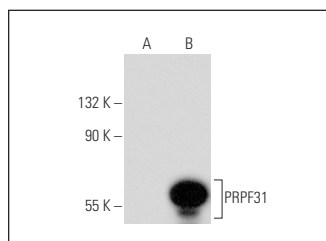
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

PRPF31 (A-6): sc-166792 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse PRPF31 expression in PRPF31 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

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.

DATA



PRPF31 (A-6): sc-166792. Western blot analysis of PRPF31 expression in non-transfected: sc-117752 (A) and mouse PRPF31 transfected: sc-127396 (B) 293T whole cell lysates.

RESEARCH USE

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