



# SZABO SCANDIC

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## Produktinformation



Forschungsprodukte & Biochemikalien



Zellkultur & Verbrauchsmaterial



Diagnostik & molekulare Diagnostik



Laborgeräte & Service

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### Lieferung & Zahlungsart

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### Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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# LRP130 (m): 293T Lysate: sc-121398

## BACKGROUND

The leucine-rich (LRR) repeat is a 20-30 amino acid motif that forms a hydrophobic  $\alpha/\beta$  horseshoe fold, allowing it to accommodate several leucine residues within a tightly packed core. All LRR repeats contain a variable segment and a highly conserved segment, the latter of which accounts for 11 or 12 residues of the entire LRR motif. Leucine-rich protein (LRP130) is a cytoplasmic mRNA-binding protein likely to be involved in the processing of mitochondrial DNA transcripts. Defects in the LRPPRC gene that encodes LRP130 result in the French-Canadian type of Leigh syndrome, a severe neurological disorder characterized by lesions in the subcortical region of the brain. LRP130 also interacts with the low-affinity receptor for leukemia inhibitory factor to produce an intracellular signal cascade.

## REFERENCES

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- Kobe, B. and Deisenhofer, J. 1995. Proteins with leucine-rich repeats. *Curr. Opin. Struct. Biol.* 5: 409-416.
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- Tsuchiya, N., et al. 2004. LRP130, a single-stranded DNA/RNA-binding protein, localizes at the outer nuclear and endoplasmic reticulum membrane, and interacts with mRNA *in vivo*. *Biochem. Biophys. Res. Commun.* 317: 736-743.
- Xu, F., et al. 2004. The role of the LRPPRC (leucine-rich pentatricopeptide repeat cassette) gene in cytochrome oxidase assembly: mutation causes lowered levels of COX (cytochrome c oxidase) I and COX III mRNA. *Biochem. J.* 382: 331-336.
- Labialle, S., et al. 2004. New invMED1 element *cis*-activates human multidrug-related MDR1 and MVP genes, involving the LRP130 protein. *Nucleic Acids Res.* 32: 3864-3876.
- Cooper, M.P., et al. 2006. Defects in energy homeostasis in Leigh syndrome French Canadian variant through PGC-1 $\alpha$ /LRP130 complex. *Genes Dev.* 20: 2996-3009.

## CHROMOSOMAL LOCATION

Genetic locus: *Lrpprc* (mouse) mapping to 17 E4.

## PRODUCT

LRP130 (m): 293T Lysate represents a lysate of mouse LRP130 transfected 293T cells and is provided as 100  $\mu$ g protein in 200  $\mu$ l SDS-PAGE buffer.

## APPLICATIONS

LRP130 (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive LRP130 antibodies. Recommended use: 10-20  $\mu$ l per lane.

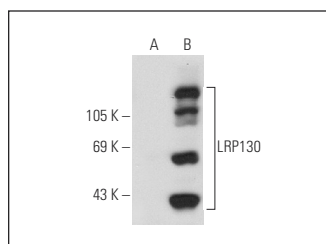
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

LRP130 (G-10): sc-166177 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse LRP130 expression in LRP130 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

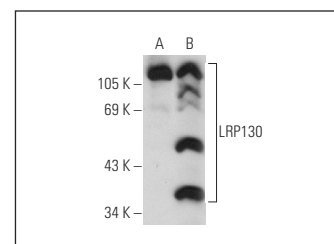
## RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended:  
 1) Western Blotting: use m-IgG $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  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



LRP130 (G-10): sc-166177. Western blot analysis of LRP130 expression in non-transfected: sc-117752 (A) and mouse LRP130 transfected: sc-121398 (B) 293T whole cell lysates.



LRP130 (F-7): sc-166178. Western blot analysis of LRP130 expression in non-transfected: sc-117752 (A) and mouse LRP130 transfected: sc-121398 (B) 293T whole cell lysates.

## 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.

## RESEARCH USE

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

## PROTOCOLS

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