

# LRP130 (H-300): sc-66844

## 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

- Hou, J., et al. 1994. Molecular cloning and expression of the gene for a major leucine-rich protein from human hepatoblastoma cells (HepG2). *In Vitro Cell. Dev. Biol. Anim.* 30A: 111-114.
- Kobe, B. and Deisenhofer, J. 1994. The leucine-rich repeat: a versatile binding motif. *Trends Biochem. Sci.* 19: 415-421.
- Kobe, B. and Deisenhofer, J. 1995. Proteins with leucine-rich repeats. *Curr. Opin. Struct. Biol.* 5: 409-416.

## CHROMOSOMAL LOCATION

Genetic locus: LRPPRC (human) mapping to 2p21; Lrpprc (mouse) mapping to 17 E4.

## SOURCE

LRP130 (H-300) is a rabbit polyclonal antibody raised against amino acids 974-1273 mapping at the C-terminus of LRP130 of human origin.

## PRODUCT

Each vial contains 200  $\mu$ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

## APPLICATIONS

LRP130 (H-300) is recommended for detection of LRP130 of human and, to a lesser extent, mouse and rat origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2  $\mu$ g per 100-500  $\mu$ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (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 LRP130 siRNA (h): sc-44734, LRP130 siRNA (m): sc-44735, LRP130 shRNA Plasmid (h): sc-44734-SH, LRP130 shRNA Plasmid (m): sc-44735-SH, LRP130 shRNA (h) Lentiviral Particles: sc-44734-V and LRP130 shRNA (m) Lentiviral Particles: sc-44735-V.

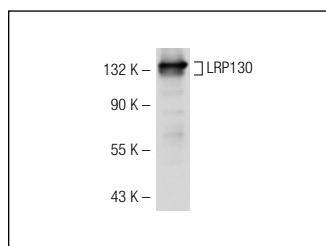
Molecular Weight of LRP130: 137 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, Hep G2 nuclear extract: sc-364819 or HeLa nuclear extract: sc-2120.

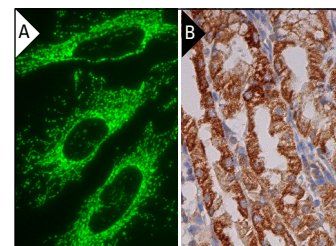
## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker<sup>™</sup> compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker<sup>™</sup> 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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz<sup>™</sup> Mounting Medium: sc-24941. 4) Immunohistochemistry: use ImmunoCruz<sup>™</sup>: sc-2051 or ABC: sc-2018 rabbit IgG Staining Systems.

## DATA



LRP130 (H-300): sc-66844. Western blot analysis of LRP130 expression in HeLa whole cell lysate.



LRP130 (H-300): sc-66844. Immunofluorescence staining of methanol-fixed HeLa cells showing mitochondrial localization (A). Immunoperoxidase staining of formalin fixed, paraffin-embedded human upper stomach tissue showing cytoplasmic staining of glandular cells (B).

## SELECT PRODUCT CITATIONS

- Schweitzer, C.J., et al. 2012. Knockdown of the cellular protein LRPPRC attenuates HIV-1 infection. *PLoS ONE* 7: e40537.
- Arun, V., et al. 2013. A novel neurofibromin (NF1) interaction with the leucine-rich pentatricopeptide repeat motif-containing protein links neurofibromatosis type 1 and the French Canadian variant of Leigh's syndrome in a common molecular complex. *J. Neurosci. Res.* 91: 494-505.

## 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.



Try **LRP130 (F-7): sc-166178** or **LRP130 (G-10): sc-166177**, our highly recommended monoclonal alternatives to LRP130 (H-300). Also, for AC, HRP, FITC, PE, Alexa Fluor<sup>®</sup> 488 and Alexa Fluor<sup>®</sup> 647 conjugates, see **LRP130 (F-7): sc-166178**.