

LMX1B (N-14): sc-21231

BACKGROUND

Nail-patella syndrome (NPS) is an autosomal dominant disorder characterized by dysplasia of finger nails, skeletal anomalies and, frequently, renal disease. NPS is caused by putative loss-of-function mutations in the transcription factor LMX1B. LMX1B belongs to the LIM-homeodomain family, members of which are known to be important for pattern formation during development. Twenty-two novel mutations may occur in the gene encoding LMX1B and the type and distribution of the mutations support the hypothesis that NPS is the result of haploinsufficiency for LMX1B. LMX1B is also necessary for normal development of the eye and in regulating dopaminergic neurogenesis and may be involved in developmental glaucoma and the aetiology of idiopathic Parkinson's disease. Specifically, LMX1B along with LIM1 control the initial trajectory of motor axons in the developing mammalian limb. In addition, LMX1B directly regulates the coordinated expression of α 3(IV) and α 4(IV) collagen required for normal glomerular basement membrane (GBM) morphogenesis, and the dysregulation of LMX1B in GBM contributes to the renal pathology and nephrosis in NPS.

CHROMOSOMAL LOCATION

Genetic locus: LMX1B (human) mapping to 9q33.3; Lmx1b (mouse) mapping to 2 B.

SOURCE

LMX1B (N-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the N-terminus of LMX1B of human origin.

PRODUCT

Each vial contains 200 μ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin. Also available as TransCruz reagent for Gel Supershift and ChIP applications, sc-21231 X, 200 μ g/0.1 ml.

Blocking peptide available for competition studies, sc-21231 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

LMX1B (N-14) is recommended for detection of LMX1B of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g 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 LMX1B siRNA (h): sc-38721, LMX1B siRNA (m): sc-38722, LMX1B shRNA Plasmid (h): sc-38721-SH, LMX1B shRNA Plasmid (m): sc-38722-SH, LMX1B shRNA (h) Lentiviral Particles: sc-38721-V and LMX1B shRNA (m) Lentiviral Particles: sc-38722-V.

LMX1B (N-14) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

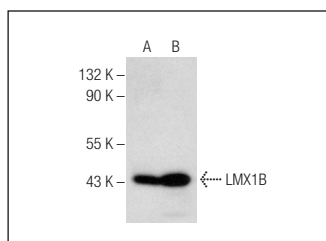
Molecular Weight of LMX1B: 42 kDa.

Positive Controls: Jurkat nuclear extract: sc-2132 or HUV-EC-C whole cell lysate: sc-364180.

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.

DATA



LMX1B (N-14): sc-21231. Western blot analysis of LMX1B expression in Jurkat nuclear extract (A) and HUV-EC-C whole cell lysate (B).

SELECT PRODUCT CITATIONS

- Arques, C.G., et al. 2007. Cell tracing reveals a dorsoventral lineage restriction plane in the mouse limb bud mesenchyme. *Development* 134: 3713-3722.
- Hoekstra, E.J., et al. 2013. LMX1B is part of a transcriptional complex with PSCP1 and PSF. *PLoS ONE* 8: e53122.

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.

PROTOCOLS

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

MONOS
Satisfaction
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Try **LMX1B (1D12): sc-293262**, our highly recommended monoclonal alternative to LMX1B (N-14).