SANTA CRUZ BIOTECHNOLOGY, INC.

LMX1B (T-25): sc-133745

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The Power to Question

BACKGROUND

Nail-patella syndrome (NPS) is an autosomal dominant disorder characterized by dyplasia 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. 22 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 alpha 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.

REFERENCES

1. Kania, A., et al. 2000. Coordinate roles for LIM homeobox genes in directing the dorsoventral trajectory of motor axons in the vertebrate limb. Cell 102: 161-173.
2. Knoers, N.V., et al. 2000. Nail-patella syndrome: identification of mutations in the LMX1B gene in Dutch families. J. Am. Soc. Nephrol. 11: 1762-1766.
3. Hamlington, J.D., et al. 2001. Twenty-two novel LMX1B mutations identified in nail patella syndrome (NPS) patients. Hum. Mutat. 18: 458.
4. Kim, B.S., et al. 2001. Targeted disruption of the myocilin gene (Myoc) suggests that human glaucoma-causing mutations are gain of function. Mol. Cell. Biol. 21: 7707-7713.
5. Morello, R., et al. 2001. Regulation of glomerular basement membrane collagen expression by LMX1B contributes to renal disease in nail patella syndrome. Nat. Genet. 27: 205-208.
6. Ramsden, D.B., et al. 2001. The aetiology of idiopathic Parkinson's disease. Mol. Pathol. 54: 369-380.

CHROMOSOMAL LOCATION

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

SOURCE

LMX1B (T-25) is a Protein A purified rabbit polyclonal antibody raised against a peptide mapping at the C-terminus of LMX1B of human origin.

PRODUCT

Each vial contains 100 µg IgG in 1.0 ml PBS with < 0.1% sodium azide, 0.1% gelatin and < 0.02% sucrose.

RESEARCH USE

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

APPLICATIONS

LMX1B (T-25) is recommended for detection of LMX1B of mouse, rat, human and zebrafish origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

LMX1B (T-25) is also recommended for detection of LMX1B in additional species, including equine, bovine and canine.

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.

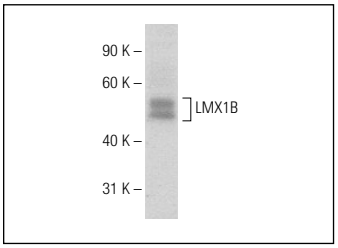
Molecular Weight of LMX1B: 42 kDa.

Positive Controls: human fetal muscle tissue extract.

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™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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).

DATA



LMX1B (T-25): sc-133745. Western blot analysis of LMX1B expression in human fetal muscle tissue extract.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.