SANTA CRUZ BIOTECHNOLOGY, INC.

Progerin (13A4D4): sc-81611

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Lamin A, also known as LMNA, is a 664 amino acid A-type nuclear lamin that is involved in nuclear stability, gene expression and chromatin structure. Alternate splicing events result in the expression of three isoforms of Lamin A, one of which is a truncated 614 amino acid protein known as Progerin (Lamin Aδ50). Progerin is created via a *de novo* silent point mutation in Lamin A and is the underlying cause of Hutchison-Gilford progeria syndrome (HGPS), a condition associated with rapidly accelerated aging characterized by cardiovascular disease and skeletal abnormalities. During interphase in normal cells, Progerin is anchored to the nuclear membrane, where it is thought to participate in mitotic events and may regulate standard aging processes. In cells affected with HGPS, however, Progerin mislocalizes into insoluble cytoplasmic aggregates and causes abnormal chromosome binucleation and segregation, thus creating the mitotic abnormality observed in HGPS.

REFERENCES

1. Zhong, N., et al. 2005. Novel progerin-interactive partner proteins hnRNP E1, EGF, Mel 18, and UBC9 interact with Lamin A/C. Biochem. Biophys. Res. Commun. 338: 855-861.
2. Glynn, M.W. and Glover, T.W. 2005. Incomplete processing of mutant Lamin A in Hutchinson-Gilford progeria leads to nuclear abnormalities, which are reversed by farnesyltransferase inhibition. Hum. Mol. Genet. 14: 2959-2969.

CHROMOSOMAL LOCATION

Genetic locus: LMNA (human) mapping to 1q22; Nes (mouse) mapping to 3 F1.

SOURCE

Progerin (13A4D4) is a mouse monoclonal antibody raised against amino acids 604-611 of Progerin of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Progerin (13A4D4) is available conjugated to agarose (sc-81611 AC), 500 µg/ 0.25 ml agarose in 1 ml, for IP; to HRP (sc-81611 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-81611 PE), fluorescein (sc-81611 FITC), Alexa Fluor® 488 (sc-81611 AF488), Alexa Fluor® 546 (sc-81611 AF546), Alexa Fluor® 594 (sc-81611 AF594) or Alexa Fluor® 647 (sc-81611 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-81611 AF680) or Alexa Fluor® 790 (sc-81611 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

STORAGE

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

APPLICATIONS

Progerin (13A4D4) is recommended for detection of Progerin of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) and immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)]; non cross-reactive with Lamin A or Lamin C.

Molecular Weight of Progerin: 70 kDa.

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. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).

SELECT PRODUCT CITATIONS

1. Jung, Y.S., et al. 2013. Loss of VHL promotes Progerin expression, leading to impaired p14/ARF function and suppression of p53 activity. Cell Cycle 12: 2277-2290.
2. Chojnowski, A., et al. 2015. Progerin reduces LAP2α-telomere association in Hutchinson-Gilford progeria. Elife 4: e07759.
3. Lo Cicero, A., et al. 2016. A high throughput phenotypic screening reveals compounds that counteract premature osteogenic differentiation of HGPS iPS-derived mesenchymal stem cells. Sci. Rep. 6: 34798.
4. Harhouri, K., et al. 2017. MG132-induced Progerin clearance is mediated by autophagy activation and splicing regulation. EMBO Mol. Med. 9: 1294-1313.
5. Messner, M., et al. 2018. Upregulation of the aging related LMNA splice variant Progerin in dilated cardiomyopathy. PLoS ONE 13: e0196739.
6. Ciazynska, M., et al. 2018. Proteins involved in cutaneous basal cell carcinoma development. Oncol. Lett. 16: 4064-4072.
7. Wu, Z., et al. 2018. Differential stem cell aging kinetics in Hutchinson-Gilford progeria syndrome and Werner syndrome. Protein Cell 9: 333-350.
8. Lo Cicero, A., et al. 2018. Pathological modelling of pigmentation disorders associated with Hutchinson-Gilford progeria syndrome (HGPS) revealed an impaired melanogenesis pathway in iPS-derived melanocytes. Sci. Rep. 8: 9112.
9. Geng, L., et al. 2018. Chemical screen identifies a geroprotective role of quercetin in premature aging. Protein Cell 10: 417-435.
10. Xu, X., et al. 2019. Progerin accumulation in nucleus pulposus cells impairs mitochondrial function and induces intervertebral disc degeneration and therapeutic effects of sulforaphane. Theranostics 9: 2252-2267.
11. Wang, F., et al. 2020. Generation of a Hutchinson-Gilford progeria syndrome monkey model by base editing. Protein Cell 11: 809-824.

RESEARCH USE

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