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Recombinant Human Progerin

REP0050 - REP0051
100µg - 50µg

Description

Progerin is a 614 amino acids protein involved in Hutchinson-Gilford progeria syndrome. Progerin is most often generated by a point mutation (C1824T) in the LMNA gene that codes for lamin A and C. This mutation activates a cryptic splice site and gives rise to a form of lamin A with a 50-amino acids internal deletion within the carboxyl-terminal domain of the protein. Approximately 80% of Hutchinson-Gilford progeria syndrome cases carry a single copy of the most common mutation, a silent point mutation, G608G (GGC > GGT), within exon 11 of LMNA gene.

Lamin A constitutes a major structural component of the lamina, a scaffold of proteins found inside the nuclear membrane of a cell; progerin does not properly integrate into the lamina, which disrupts the scaffold structure and leads to significant disfigurement of the nucleus, characterized by a lobular shape.

Product type

Recombinant Protein

Peptide

Recombinant full-length protein: Human progerin – Accession number: **AAR29466.1**

Sequence

MAHHHHHVGVTGNSDDDDKSPDMETPSQRRATRSGAQASSTPLSPTRITRLQEKEDLQELNDR LAVYIDRVRS
LETENAGLRLRITESEEVVSREVSIGIKAAEAEELGDARKTLDSVAKERARLQLELSKVREEFKELKARNTKKEGDLI
AAQARLKDLEALLNSKEAALSTALSEKRTLEGELHDLRGQVAKLEAALGEAKKQLQDEMLRRVDAENRLQTMKEE
LDFQKNIYSEELRETKRRHETRLVEIDNGKQREFESRLADALQELRAQHEDQVEQYKKELEKTYSAKLDNARQSA
ERNSNLVGAHHEELQQRIRIDSLSAQLSQLQKQLAAKEAKLRDLEDLSLARERDTSRRLAEEKEREMAEMRARMQ
QQLDEYQELLDIKLALDMEIHAYRKLLGEEERLRLSPSPTSQRSRGRASSHSSQTQGGGSVTKKRKLESTESRSS
FSQHARTSGRVAVEEVDEEGKFVRLRNKSNEDQSMGNWQIKRQNGDDPLLTYRFPKFTLKAGQVVTIWAAGAG
ATHSPPTDLVWKAQNTWGCNSLRTALINSTGEEVAMRKLVRSVTVVEDDEDEDGDDLLHHHHGSHCSSSGDPA
EYNLRSRTLVCCTCGQPADKASASGGAQ[]SPQNC SIM

Escherichia coli

Expression system

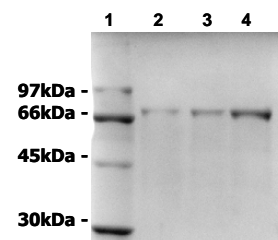
Tested by

SDS Page, Western Blotting

Purity

> 90% pure estimated by SDS-PAGE (EU Ph. 5.0 § 2.5.31)

Purified recombinant human progerin protein (lane 1, molecular weight standard; lane 2, 1µg; lane 3, 3µg; lane 4, 5µg) was separated by SDS-PAGE (10% polyacrylamide) and stained with Coomassie Blue.



Form

Liquid

Storage buffer

20mM Tris-HCl pH 7.5, 0,5M NaCl, 1mM DTT, 1,5mM EDTA, 20%(v/v) glycerol.

Storage instructions

Shipped on dry ice. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.