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## Recombinant Rat Progerin

**REP0047**  
**100µg**

### Description

Progerin is a 614 amino acid 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: rat progerin

### Sequence

METPSQRRATRSGAQASSTPLSPTRITRLQEKEDLQELNDR LAVYIDRVRSLETENAGLRLRITESEEVVSREVS GIKAAAYEA  
ELGDARKTLDSVAKERARLQLELSKVREEFKELKARNTKKEGDL LAAQARLKDLEALLNSKEAALSTALSEKRTLEGELHDLR  
GQVAKLEAALGEAKKQLQDEMLRRVDAENRLQTLKEELDFQKN IYSEELRETKRRHETRLVEIDNGKQREFESRLADALQEL  
RAQHEDQVEQYKKELEKTYSAKLDNARQSAERNSNLVGA AHEELQQSRIRIDSLSAQLSQLQKLAAKEAKLRDLED SLAR  
ERDTSRLLAEKEREMAEMRARMQQQLDEYQELLDIKLALD MEIHAYRKLLEGEERLRLSPSPTSQRSRGRASSHSSQSQ  
GGGSVTKKRKLESSESRSFSQHARTSGRVAVEEVDEEGKFV RLRNKSNEQSMGNWQIRRNQNGDDPLMTYRFPPKFTLK  
AGQVVTIWASGAGATHSPPTDLVWKAQNTWGCSSSLRTAL INATGEEVAMRKLVRSLTMVEDDEDEDGDDLLHHHHGSH  
CSSSGDPAEYNLRSRTVLCGTGCGPADKASASGSGAQV[ ]SSQNCSIM

### Expression system

*Escherichia coli*

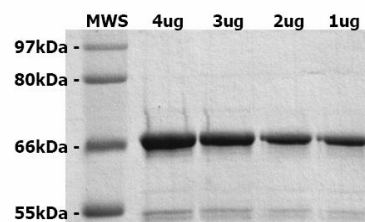
### Tested by

SDS Page, Western Blotting

### Purity

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

Purified recombinant full-length rat progerin protein was separated by SDS-PAGE (8% polyacrylamide) and stained with Coomassie Blue.



### Form

Liquid

### Storage buffer

20mM Tris-HCl pH 7.5, 0,3M NaCl, 1mM DTT, 1,5mM EDTA, 10%(v/v) glycerol.

### Storage instructions

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