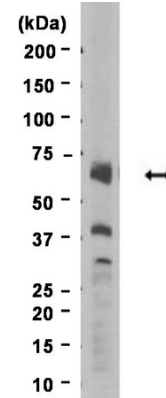


Monoclonal Antibody to Progerin - Purified

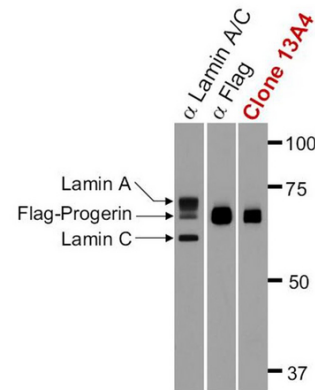
Alternate names:	CMD1A, EMD2, FPL, FPLD, LDP1, LFP, LMN1, LMNA, LMNC
Catalog No.:	AM50371PU-N
Quantity:	0.1 mg
Concentration:	1.0 mg/ml (lot-specific)
Background:	<p>Progerin is a truncated version of lamin A protein involved in Hutchinson-Gilford progeria syndrome. Progerin is most often generated by a mutation (C1824T) in the lamin A gene, LMNA. This mutation activates a cryptic splice site and gives rise to a form of lamin A with a deletion of 50 amino acids. 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. Researchers have shown that progerin activates genes that regulate stem cell differentiation via the Notch signaling pathway. Progerin, which has been linked to normal aging, is produced in healthy individuals via "sporadic use of the cryptic splice site.</p>
Uniprot ID:	Q6UYC3
NCBI:	9606
Host / Isotype:	Mouse / IgG1
Recommended Isotype Controls:	SM10P (for use in human samples), AM03095PU-N
Clone:	13A4
Immunogen:	KLH-conjugated synthetic peptide corresponding to the amino acids 604-611 of progerin (lamin A/C).
Format:	State: Liquid purified IgG fraction from Ascites Purification: Protein G Chromatography Buffer System: 0.1 M Tris-Glycine (pH7.4), 150 mM NaCl, 0.05% sodium azide.
Applications:	Immunoprecipitation. Western Blot. Immunocytochemistry. ELISA. Other applications not tested. Optimal dilutions are dependent on conditions and should be determined by the user.
Specificity:	Recognizes human progerin, Mr ~70 kDa. Does not recognize Lamin A or Lamin C Human. Other species not tested.
Storage:	Store undiluted at 2-8°C. Shelf life: one year from despatch.

Pictures:

Western Blot Analysis:
 Transfected HeLa cell lysate expressing progerin was resolved by electrophoresis, transferred to PVDF and probed with anti-progerin (1 µg/ml). Proteins were visualized using a goat anti-mouse secondary antibody conjugated to HRP and a chemiluminescence detection system.



Whole cell lysate of HeLa cells ectopically expressing Flag-tagged human progerin (Protein Accession #AAR29466) was resolved by Western blot (10% PAGE) and transferred to PVDF. The membrane was cut into strips and each strip was incubated separately with the following antibodies: Lane 1: anti-Lamin A/C; Lane 2: anti-Flag-tag antibody (Sigma, 1:5000); Lane 3: Anti-Progerin, clone 13A4. Anti-Progerin clone 13A4 specifically detects human progerin by immunoblotting. Images generously provided by Dr. Egon Ogris, University of Vienna.



Whole cell lysate of T98G cells treated with FTI-277 (EMD Biosciences), a farnesyl transferase inhibitor that causes accumulation of unprocessed Lamin A (Prelamin A), was resolved by Western blot and transferred to PVDF. The membrane was cut into strips and each strip was incubated separately with the following antibodies: Lane 1: anti-Lamin A/C; Lane 2: anti-Pre-Lamin A (Santa Cruz); Lane 3: Anti-Progerin, clone 13A4. Anti-Progerin clone 13A4 does not cross-react with prelamin A. Images generously provided by Dr. Egon Ogris, University of Vienna.

