Product Information

Monoclonal Anti-Progerin

Clone 13A4, produced in Mouse
Purified Immunoglobulin

SAB4200272

Product Description

Monoclonal Anti-Progerin (mouse IgG1 isotype) is derived from the hybridoma 13A4 produced by the fusion of mouse myeloma cells and splenocytes from BALB/c mice immunized with a synthetic peptide corresponding to a fragment of progerin (mutated Lamin A/C) (GeneID: 4000), conjugated to KLH. The isotype is determined by ELISA using Mouse Monoclonal Antibody Isotyping Reagents, (Cat. No. ISO2). The antibody is purified from culture supernatant of hybridoma cells grown in a bioreactor.

Monoclonal Anti-Progerin recognizes human progerin and does not recognize Lamin A or Lamin C. The antibody may be used in several immunochemical techniques including immunoblotting (~ 70 kDa), immunoprecipitation, and immunofluorescence.[1]

The nuclear lamina is a meshwork of intermediate filaments that underlies the inner face of the nuclear envelope.[2] The major components of the nuclear lamina are the lamins that may be classified into two types, A and B. Both A- and B- type lamins are characterized by an α-helical rod domain to enable assembly into filaments, a nuclear localization sequence, and a carboxy-terminal CAAX box isoprenylation sequence for nuclear membrane targeting.[3] A-type lamins, lamin A and lamin C, are products of a single gene, *LMNA,* which are produced by alternative splicing.[4] Prelamin A, the precursor of Lamin A, is farnesylated at its carboxy terminus after synthesis, and the last 18 amino acids, which contain the farnesyl group, are removed by the metalloprotease Zmpste24, producing the mature Lamin A.[5] Mutations in Lamin A have been linked to a variety of rare human diseases including muscular dystrophy, lipodystrophy, cardiomyopathy, neuropathy and progeroid syndromes (collectively termed laminopathies) and to premature aging (Hutchinson-Gilford progeria syndrome).[6,7]

The most common form of progeria is caused by the expression of a mutant lamin A protein, progerin, which arises from defective maturation of prelamin A. Progerin is generated by a 1824C>T mutation (G608G), which activates a cryptic splice site resulting in the expression of lamin A with a deletion of 50 amino acids near its carboxy terminus. As a result, progerin lacks the target sequence for Zmpste24 and remains constitutively farnesylated. Accumulation of farnesylated progerin causes severe abnormalities of the nuclear envelope, cellular senescence, and genomic instability.[8,9]

Reagent

Supplied as a solution in 0.01 M phosphate buffered saline, pH 7.4, containing 15 mM sodium azide as a preservative.

Antibody concentration: ~ 1.0 mg/mL

Precautions and Disclaimer

For research use only. Not for drug, household, or other uses. Please consult the Safety Data Sheet for information regarding hazards and safe handling practices.

Storage/Stability

For continuous use, store at 2–8 °C for up to one month. For extended storage, freeze at –20 °C in working aliquots. Repeated freezing and thawing, or storage in "frost-free" freezers, is not recommended. If slight turbidity occurs upon prolonged storage, clarify the solution by centrifugation before use. Working dilution samples should be discarded if not used within 12 hours.

Product Profile

Immunoblotting: a working concentration of 2.5-5.0 µg/mL is recommended using whole extracts of HeLa cells overexpressing human progerin.

Note: In order to obtain best results in various techniques and preparations, it is recommended to determine optimal working dilutions by titration.

References

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