

Product Information

Anti-Lamin A/C R48W Antibody, Mouse Monoclonal

Clone 5H8-B4, Purified from Hybridoma Cell Culture

SAB4200422

Product Description

Anti-Lamin A/C R482W (mouse IgG1 isotype) is derived from the hybridoma 5H8-B4 produced by the fusion of mouse myeloma cells and splenocytes from BALB/c mice immunized with a recombinant protein fragment containing the R482W substitution of human Lamin A/C (GeneID: 4000).¹ 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. Anti-Lamin A/C R482W recognizes a human Lamin A/C recombinant protein containing the R482W substitution. This antibody does not recognize the Lamin A/C wild-type protein. The antibody may be used in several immunochemical techniques including immunoblotting, immunoprecipitation and immunofluorescence.¹

Lamin A is a structural protein of the nuclear lamina. 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, resulting in proteins of 664 and 572 amino acids, respectively. ⁴ The first 566 amino acids of Lamin A and C are identical. Prelamin A, the precursor of Lamin A, has 98 unique amino acids and is farnesylated at its carboxy terminus after synthesis. The last 18 amino acids, which contain the farnesyl group, are removed by an endoproteolytic cleavage, producing the mature Lamin A.⁵

Lamins are expressed in most somatic cells. They interact with integral proteins of the inner membrane of the nuclear envelope, such as LAPs 1 and 2 (lamina- associated polypeptides), LBR (Lamin B receptor) and emerin.⁶ They also interact with chromatin and nuclear pore complexes.⁵ Lamin A is cleaved into a 47 kDa fragment during apoptosis. This cleavage seems to be essential for chromatin condensation and nuclear disassembly in apoptosis.^{3,7} Mutations in Lamin A and C 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).^{8,9} Most diseases arise from dominant, missense mutations. Lamin A/C R482W antibody is a critical tool for basic and clinical research of the mutant A-type lamins in primary patient cells.

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 R&D 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 0.1-0.2 µg/mL is recommended using a recombinant protein encoding a fragment of human Lamin A/C containing the R482W substitution.

Note: In order to obtain the best results using various techniques and preparations, we recommend determining optimal working dilutions by titration.

References

1. Roblek, M., et al., *PLoS ONE*, **5**: e10604 (2010).
2. Aeby, U., et al., *Nature*, **323**: 560-564 (1986).
3. Rao, L., et al., *J. Cell Biol.*, **135**: 1441-1455 (1996).
4. Lloyd, D.J., et al., *Hum. Mol. Gen.*, **11**: 769-777 (2002).
5. Worman, H.J. and Courvalin, J.C., *Trends Cell Biol.*, **12**: 591-598 (2002).
6. Schirmer, E.C., et al., *J. Cell Biol.*, **153**: 479-489 (2001).
7. Ruchaud, S., et al., *EMBO J.*, **21**: 1967-1977 (2002).
8. Worman, H.J., and Courvalin, J.C., *J. Clin. Invest.*, **113**: 349-351 (2004).
9. Plasilova, M., et al., *J. Med. Genet.*, **41**: 609-614 (2004).

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