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Product Information

Anti-DKC1
produced in rabbit, affinity isolated antibody

Catalog Number **SAB4200611**

Product Description

Anti-DKC1 is produced in rabbit using as immunogen a peptide corresponding to the N-terminal region of human DKC1 (GeneID: 1736), conjugated to KLH. The corresponding sequence is identical in monkey, bovine, pig and dog and differs by a single amino acid in mouse and rat. The antibody is affinity-purified using the immunizing peptide immobilized on agarose.

Anti-DKC1 recognizes human and mouse DKC1. The antibody may be used in various immunochemical techniques including immunoblotting (~57 kDa). Detection of the DKC1 band by immunoblotting is specifically inhibited by the immunizing peptide.

Dyskerin is a nucleolar protein encoded by the *DKC1* gene. This gene is a member of the H/ACA snoRNPs (small nucleolar ribonucleoproteins) gene family. snoRNPs are involved in various aspects of rRNA processing and modification and have been classified into two families: C/D and H/ACA. The H/ACA snoRNPs also include the NOLA1, 2 and 3 proteins. Dyskerin and the three NOLA proteins localize to the dense fibrillar components of nucleoli and to coiled (Cajal) bodies in the nucleus. Both 18S rRNA production and rRNA pseudouridylation are impaired if any one of the four proteins is depleted. These four H/ACA snoRNP proteins are also components of the telomerase complex. Dyskerin is related to the Saccharomyces cerevisiae Cbf5p and Drosophila melanogaster Nop60B proteins. The *DKC1* gene lies in a tail-to-tail orientation with the palmitoylated erythrocyte membrane protein gene and is transcribed in a telomere to centromere direction. Both nucleotide substitutions and single trinucleotide repeat polymorphisms have been found in this gene. Mutations in the *DKC1* gene cause X-linked dyskeratosis congenita, a disease resulting in reticulate skin pigmentation, mucosal leukoplakia, nail dystrophy, and progressive bone marrow failure in most cases. Mutations in this gene also cause Hoyeraal-Hreidarsson syndrome, which is a more severe form of dyskeratosis congenita. The overexpression of dyskerin has been reported in several cancers, including neuroblastoma, lymphoma, melanoma, breast cancer, prostate cancer, colorectal cancer, ovarian carcinoma and hepatocellular carcinoma.[1-4]

Reagent

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

Antibody Concentration: ~ 1.0 mg/mL

Precautions and Disclaimer

This product is for R&D use only, not for drug, household, or other uses. Please consult the Material 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 in working aliquots. Repeated freezing and thawing 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 0.5-1.0 µg/mL is recommended using whole extracts of mouse Hepa1-6 cells or nuclear extract of HeLa cells.

Note: In order to obtain best results in different techniques and preparations we recommend determining optimal working concentration by titration test.

References

1. Mochizuki, Y., et al., *Proc. Natl. Acad. Sci. USA*, **101**, 10756-10761 (2004).
2. Jack, K., et al., *Mol. Cell*, **44**, 660-666 (2011).
3. Egan, E.D., and Collins, K., *Mol. Cell. Biol.*, **32**, 2428-2439 (2012).
4. Liu, B., et al., *PloS One*, **7**, e43147 (2012).

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