RecomAb™

DKC1 recombinant monoclonal antibody, clone R03-6X5

Catalog # RAB05274 Size 100 uL

Specification

Product Description	Rabbit recombinant monoclonal antibody raised against human DKC1.
Antibody Species	Rabbit
Immunogen	Original antibody is raised against recombinant protein corresponding to human DKC1
Theoretical MW (kDa)	Calculated MW: 58 kD
Reactivity	Human
Form	Liquid
Purification	Affinity purification
Isotype	IgG
Recommend Usage	Immunofluorescence (1/50-1/200) Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections)(1/50-1/100) Western Blot (1/500-1/1000) The optimal working dilution should be determined by the end user.
Storage Buffer	In PBS, 150 mM NaCl, pH 7.4 (50% glycerol and 0.02% Sodium azide)
Storage Instruction	Store at 4°C. For long term storage store at -20°C. Aliquot to avoid repeated freezing and thawing.
Note	This product contains sodium azide: a POISONOUS AND HAZARDOUS SUBSTANCE which shoul d be handled by trained staff only.

Applications

- Western Blot
- Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections)

- Immunocytochemistry
- Immunofluorescence

Gene Info — DKC1

Entrez GeneID	1736
Gene Name	DKC1
Gene Alias	CBF5, DKC, FLJ97620, NAP57, NOLA4, XAP101
Gene Description	dyskeratosis congenita 1, dyskerin
Omim ID	300126 300240 305000
Gene Ontology	Hyperlink
Gene Summary	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 cl assified into two families: C/D and H/ACA. The H/ACA snoRNPs also include the NOLA1, 2 and 3 proteins. The protein encoded by this gene and the three NOLA proteins localize to the dense fi brillar components of nucleoli and to coiled (Cajal) bodies in the nucleus. Both 18S rRNA producti on and rRNA pseudouridylation are impaired if any one of the four proteins is depleted. These fou r H/ACA snoRNP proteins are also components of the telomerase complex. The protein encoded by this gene is related to the Saccharomyces cerevisiae Cbf5p and Drosophila melanogaster No p60B proteins. The gene lies in a tail-to-tail orientation with the palmitoylated erythrocyte membra ne protein gene and is transcribed in a telomere to centromere direction. Both nucleotide substitut ions and single trinucleotide repeat polymorphisms have been found in this gene. Mutations in this 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. Mutatio ns in this gene also cause Hoyeraal-Hreidarsson syndrome, which is a more severe form of dyske ratosis congenita. Two transcript variants encoding different isoforms have been found for this ge ne. [provided by RefSeq
Other Designations	H/ACA ribonucleoprotein complex subunit 4|OTTHUMP00000026046|cbf5p homolog|dyskerin|no pp140-associated protein of 57 kDa|nucleolar protein family A member 4|snoRNP protein DKC1

Disease

- Anemia