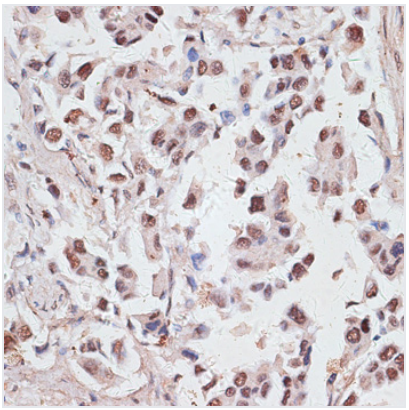
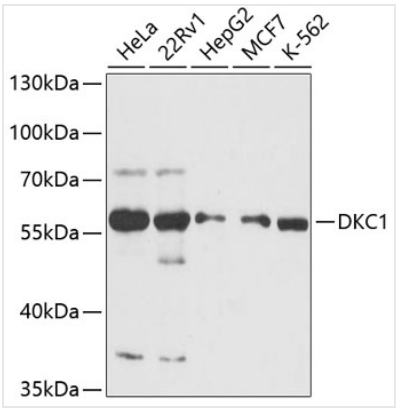
# DKC1 Antibody – Rabbit Polyclonal

## PRODUCT OVERVIEW

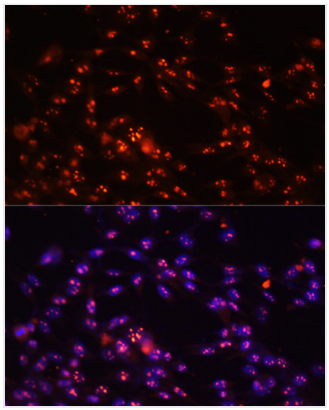
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|---|---|
| **Product Name** | DKC1, Polyclonal Antibody |
| **Full Product Name** | DKC1 Rabbit Polyclonal Antibody |
| **Product Gene Name** | anti-DKC1 antibody |
| **Product Synonym Names** | DKC1; CBF5; DKC; DKCX; NAP57; NOLA4; XAP101 |
| **Clonality** | Polyclonal |
| **Isotype** | IgG |
| **Host** | Rabbit |
| **Species Reactivity** | Human, Mouse |
| **Form/Format** | Buffer: PBS with 0.02% sodium azide, 50% glycerol, pH7.3. |
| **Purity/Purification** | Affinity purification |
| **Preparation and Storage** | Store at -20 degree C. Avoid freeze/thaw cycles. |
| **Molecular Weight** | 57,674 Da |
| **Chromosome Location** | Chromosome: X; NC_000023.10 (153991031..154005964). Location: Xq28 |
| **OMIM** | 300126 |
| **3D Structure** | ModBase 3D Structure for O60832 |
| **Immunogen** | Recombinant fusion protein of human DKC1 (NP_001354.1). |
| **Product Categories/Family for anti-DKC1 antibody** | Primary Antibody |
| **Applications Tested/Suitable for anti-DKC1 antibody** | Western Blot (WB), Immunohistochemistry (IHC), Immunofluorescence (IF) |
| **Application Notes for anti-DKC1 antibody** | WB: 1:500-1:2000<br>IHC: 1:50-1:100<br>IF: 1:50-1:100 |
| **Related Product Information for anti-DKC1 antibody** | This gene functions in two distinct complexes. It plays an active role in telomerase stabilization and maintenance, as well as recognition of snoRNAs containing H/ACA sequences which provides stability during biogenesis and assembly into H/ACA small nucleolar RNA ribonucleoproteins (snoRNPs). This gene is highly conserved and widely expressed, and may play additional roles in nucleo-cytoplasmic shuttling, DNA damage response, and cell adhesion. Mutations have been associated with X-linked dyskeratosis congenita. Alternative splicing results in multiple transcript variants. |
| **Other Notes** | Small volumes of anti-DKC1 antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply. |
| **Searchable Terms for DKC1 purchase** | MBS9464721 is an antibody targeting DKC1. Also known as DKC1 Rabbit Polyclonal Antibody. Reactive in Human, Mouse. Host: Rabbit. |



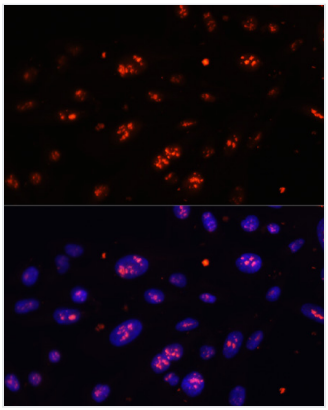
**Immunohistochemistry (IHC) of anti-DKC1 antibody**
Immunohistochemistry of paraffin-embedded human lung cancer using DKC1 at dilution of 1:100 (40x lens).




**Western Blot (WB) of anti-DKC1 antibody**
Western blot analysis of extracts of various cell lines, using DKC1 at 1:3000 dilution.



**Immunofluorescence (IF) of anti-DKC1 antibody**
Immunofluorescence analysis of NIH-3T3 cells using DKC1 at dilution of 1:100. Blue: DAPI for nuclear staining.



**Immunofluorescence (IF) of anti-DKC1 antibody**
Immunofluorescence analysis of U-2 OS cells using DKC1 at dilution of 1:100. Blue: DAPI for nuclear staining.

## ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for DKC1. It may not necessarily be applicable to this product.

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| **NCBI GI #** | 4503337 |
| **NCBI GeneID** | 1736 |
| **NCBI Accession #** | NP_001354.1 |
| **UniProt Primary Accession #** | O60832 |
| **UniProt Secondary Accession #** | O43845; Q96G67; Q9Y505; F5BSB3 |
| **UniProt Related Accession #** | O60832 |
| **GenBank Nucleotide #** | NM_001363.3 |
| **NCBI Official Full Name** | H/ACA ribonucleoprotein complex subunit 4 isoform 1 |
| **NCBI Official Synonym Full Names** | dyskeratosis congenita 1, dyskerin |
| **NCBI Official Symbol** | DKC1 |
| **NCBI Official Synonym Symbols** | DKC; CBF5; DKCX; NAP57; NOLA4; XAP101 |
| **UniProt Protein Name** | H/ACA ribonucleoprotein complex subunit 4 |
| **UniProt Synonym Protein Names** | CBF5 homolog; Dyskerin; Nopp140-associated protein of 57 kDa; Nucleolar protein NAP57; Nucleolar protein family A member 4; snoRNP protein DKC1 |

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| **UniProt Gene Name** | DKC1 |
| **UniProt Synonym Gene Names** | NOLA4 |
| **UniProt Entry Name** | DKC1_HUMAN |
| **NCBI Protein Information** | H/ACA ribonucleoprotein complex subunit 4; CBF5 homolog; cbf5p homolog; snoRNP protein DKC1; nucleolar protein NAP57; nucleolar protein family A member 4; nopp140-associated protein of 57 kDa |
| **NCBI Summary for DKC1** | 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. The protein encoded by this gene 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. The protein encoded by this gene is related to the Saccharomyces cerevisiae Cbf5p and Drosophila melanogaster Nop60B proteins. The 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 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. Mutations in this gene also cause Hoyeraal-Hreidarsson syndrome, which is a more severe form of dyskeratosis congenita. Two transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, Feb 2013] |
| **UniProt Comments for DKC1** | dyskerin: Isoform 1: Required for ribosome biogenesis and telomere maintenance. Probable catalytic subunit of H/ACA small nucleolar ribonucleoprotein (H/ACA snoRNP) complex, which catalyzes pseudouridylation of rRNA. This involves the isomerization of uridine such that the ribose is subsequently attached to C5, instead of the normal N1. Each rRNA can contain up to 100 pseudouridine ('psi') residues, which may serve to stabilize the conformation of rRNAs. Also required for correct processing or intranuclear trafficking of TERC, the RNA component of the telomerase reverse transcriptase (TERT) holoenzyme. Defects in DKC1 are a cause of dyskeratosis congenita X-linked recessive (XDKC). XDKC is a rare, progressive bone marrow failure syndrome characterized by the triad of reticulated skin hyperpigmentation, nail dystrophy, and mucosal leukoplakia. Early mortality is often associated with bone marrow failure, infections, fatal pulmonary complications, or malignancy. Defects in DKC1 are the cause of Hoyeraal-Hreidarsson syndrome (HHS). HHS is a multisystem disorder affecting males and is characterized by aplastic anemia, immunodeficiency, microcephaly, cerebellar hypoplasia, and growth retardation. Belongs to the pseudouridine synthase TruB family. 2 isoforms of the human protein are produced by alternative splicing.<br><br>Protein type: Isomerase; RNA processing; RNA-binding; Lyase; EC 5.4.99.-; Nucleolus<br><br>Chromosomal Location of Human Ortholog: Xq28<br><br>Cellular Component: nucleoplasm; Cajal body; cytoplasm; telomerase holoenzyme complex; nucleolus; nucleus<br><br>Molecular Function: telomerase activity; protein binding; RNA binding; pseudouridine synthase activity<br><br>Biological Process: RNA processing; cell proliferation; pseudouridine synthesis; telomere maintenance via telomerase; telomere maintenance; rRNA processing<br><br>Disease: Dyskeratosis Congenita, X-linked |
| **Research Articles on DKC1** | **1.** We describe an African American family with Hoyeraal-Hreidarrson syndrome (HHS) in which 2 TERT mutations (causing P530L and A880T amino acid changes) and two in the DKC1 variants (G486R and A487A) were segregating. |
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