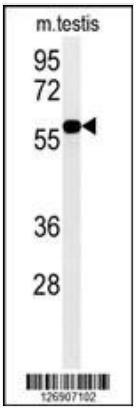
DKC1 Antibody – Rabbit Polyclonal

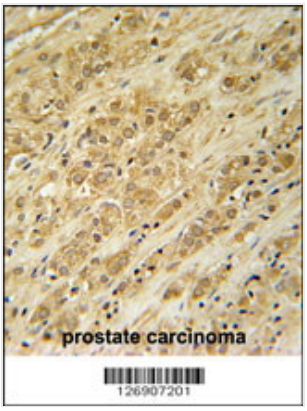
PRODUCT OVERVIEW

Product Name	DKC1, Polyclonal Antibody
Full Product Name	DKC1, ID (DKC1, NOLA4, H/ACA ribonucleoprotein complex subunit 4, CBF5 homolog, Dyskerin, Nopp140-associated protein of 57kD, Nucleolar protein NAP57, Nucleolar protein family A member 4, snoRNP protein DKC1) (Azide free) (HRP)
Product Gene Name	anti-DKC1 antibody
Product Synonym Names	DKC1; NOLA4; H/ACA ribonucleoprotein complex subunit 4; CBF5 homolog; Dyskerin; Nopp140-associated protein of 57kD; Nucleolar protein NAP57; Nucleolar protein family A member 4; snoRNP protein DKC1
Clonality	Polyclonal
Isotype	IgG
Host	Rabbit
Species Reactivity	Human, Mouse
Form/Format	Supplied as a liquid in PBS, pH 7.2. No preservative added. Labeled with Horseradish Peroxidase (HRP).
Purity/Purification	Purified by Protein A Affinity Chromatography.
Preparation and Storage	Store product at 4 degree C if to be used immediately within two weeks. For long-term storage, aliquot to avoid repeated freezing and thawing and store at -20 degree C. Aliquots are stable at -20 degree C for 12 months after receipt. Dilute required amount only prior to immediate use. Further dilutions can be made in assay buffer. Note: Sodium azide is a potent inhibitor of peroxidase and should not be added to HRP conjugates. For maximum recovery of product, centrifuge the original vial after thawing and prior to removing the cap.
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	DKC1 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 192-221 amino acids from the Central region of human DKC1.
Conjugate	HRP
Note	Preservative Free
Product Categories/Family for anti-DKC1 antibody	Antibodies; Nuclear Proteins
Applications Tested/Suitable for anti-DKC1 antibody	Immunohistochemistry (IHC), ELISA (EIA), Western Blot (WB)
Application Notes for anti-DKC1 antibody	IHC: 1:50-100 ELISA: 1:1,000 WB: 1:100-500 Applications are based on unconjugated antibody.
Related Product Information for anti-DKC1 antibody	MBS6292249 is an antibody targeting DKC1. Also known as DKC1, ID (DKC1, NOLA4, H/ACA ribonucleoprotein complex subunit 4, CBF5 homolog, Dyskerin, Nopp140-associated protein of 57kD, Nucleolar protein NAP57, Nucleolar protein family A member 4, snoRNP protein DKC1) (Azide free) (HRP). Reactive in Human, Mouse. Host: Rabbit.

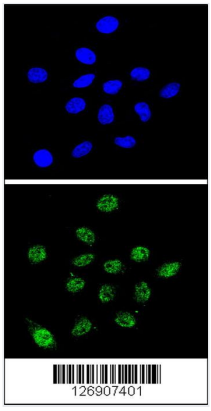
Product References and Citations for anti-DKC1 antibody	Rostamiani, K., et al. Blood Cells Mol. Dis. 44 (2), 88 (2010): Grozdanov, P.N., et al. Hum. Mol. Genet. 18(23):4546-4551(2009). Ashbridge, B., et al. Biochemistry 48(46):10858-10865(2009). Sieron, P., et al. Br. J. Cancer 101(8):1410-1416(2009).
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.



Western Blot (WB) of anti-DKC1 antibody
DKC1 Antibody (Center) western blot analysis in mouse testis tissue lysates (35ug/lane).This demonstrates the DKC1 antibody detected DKC1 protein (arrow).



Immunohistochemistry (IHC) of anti-DKC1 antibody
DKC1 Antibody (Center) immunohistochemistry analysis in formalin fixed and paraffin embedded human prostate carcinoma followed by peroxidase conjugation of the secondary antibody and DAB staining. This data demonstrates the use of the DKC1 Antibody (Center) for immunohistochemistry. Clinical relevance has not been evaluated.



Immunofluorescence (IF) of anti-DKC1 antibody
Confocal immunofluorescent analysis of DKC1 Antibody (Center) with 293 cell followed by Alexa Fluor 488-conjugated goat anti-rabbit IgG (green). DAPI was used to stain the cell nuclear (blue).

ADDITIONAL DETAILS

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

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
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. Protein type: Isomerase; RNA processing; RNA-binding; Lyase; EC 5.4.99.-; Nucleolus Chromosomal Location of Human Ortholog: Xq28 Cellular Component: nucleoplasm; Cajal body; cytoplasm; telomerase holoenzyme complex; nucleolus; nucleus Molecular Function: telomerase activity; protein binding; RNA binding; pseudouridine synthase activity Biological Process: RNA processing; cell proliferation; pseudouridine synthesis; telomere maintenance via telomerase; telomere maintenance; rRNA processing 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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