

Dyskerin (D6N4K) Rabbit mAb

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Applications:	Reactivity:	Sensitivity:	MW (kDa):	Source/Isotype:	UniProt ID:	Entrez-Gene Id:
W	H M Mk	Endogenous	60	Rabbit IgG	#O60832	1736

Product Usage Information**Application**

Western Blotting

Dilution

1:1000

Storage

Supplied in 10 mM sodium HEPES (pH 7.5), 150 mM NaCl, 100 µg/ml BSA, 50% glycerol and less than 0.02% sodium azide. Store at -20°C. Do not aliquot the antibody.

Specificity/Sensitivity

Dyskerin (D6N4K) Rabbit mAb recognizes endogenous levels of total dyskerin protein.

Source / Purification

Monoclonal antibody is produced by immunizing animals with a synthetic peptide corresponding to residues surrounding Phe91 of human dyskerin protein.

Background

H box/ACA-motif small nucleolar RNAs (snoRNAs) guide snoRNA proteins (snoRNPs) to uridine residues on rRNA for the conversion to pseudouridine (1). These H/ACA snoRNPs consist of four highly conserved proteins including GAR1, NHP2, NOP10, and the catalytic component dyskerin (1-3). The core snoRNPs also bind to mammalian telomerase RNA, which contains a H/ACA-like motif in the 3' domain. This binding results in the maintenance of telomerase levels and activity (4). Defects in the snoRNPs can lead to dyskeratosis congenita, a rare, x-linked disorder characterized by a failure of the bone marrow and an increased tumor risk (5,6). Mutations in the *dyskerin* gene can cause defects in translation of mRNAs containing internal ribosome entry sites (IRESs), which include mRNAs to tumor suppressors p27 and p53 and anti-apoptotic factors Bcl-xL and XIAP (7).

Background References

1. Lafontaine, D.L. et al. (1998) *Genes Dev* 12, 527-37.
2. Henras, A. et al. (1998) *EMBO J* 17, 7078-90.
3. Watkins, N.J. et al. (1998) *RNA* 4, 1549-68.
4. Mitchell, J.R. et al. (1999) *Nature* 402, 551-5.
5. Heiss, N.S. et al. (1998) *Nat Genet* 19, 32-8.
6. Knight, S.W. et al. (1999) *Am J Hum Genet* 65, 50-8.
7. Yoon, A. et al. (2006) *Science* 312, 902-6.

Species Reactivity

Species reactivity is determined by testing in at least one approved application (e.g., western blot).

Western Blot Buffer

IMPORTANT: For western blots, incubate membrane with diluted primary antibody in 5% w/v BSA, 1X TBS, 0.1% Tween@ 20 at 4°C with gentle shaking, overnight.

Applications Key

W: Western Blotting

Cross-Reactivity Key

H: Human **M:** Mouse **Mk:** Monkey

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