SANTA CRUZ BIOTECHNOLOGY, INC.

Dyskerin (H-300): sc-48794

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Dyskerin (NAP57) associates with the chaperone protein Nopp140 and forms a small ribonucleoprotein particle with GAR1 (NOLA1), NHP2 (NOLA2) and Nop10 for the isomerization of uridine to pseudouridine. GAR1, NHP2 and Dyskerin localize to the dense fibrillar component of the nucleolus and in nuclear Cajal bodies. The Dyskerin gene maps to chromosome Xq28. Missense mutations in the Dyskerin gene interfere with normal nuclear localization of Dyskerin and cause Dyskeratosis congenita (DKC). DKC is a rare, X-linked bone marrow disorder characterized by cutaneous hyperpigmentation, dystrophy of the nails, atrophy of the testicles and leukoplakia of the oral mucosa. The GAR1 gene maps to chromosome 4q25. The NHP2 gene maps to chromosome 5q35.3 and encodes a 155 amino acid protein.

CHROMOSOMAL LOCATION

Genetic locus: DKC1 (human) mapping to Xq28; Dkc1 (mouse) mapping to X A7.3.

SOURCE

Dyskerin (H-300) is a rabbit polyclonal antibody raised against amino acids 171-470 mapping within an internal region of Dyskerin of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Available as TransCruz reagent for Gel Supershift and ChIP applications, sc-48794 X, 200 µg/0.1 ml.

APPLICATIONS

Dyskerin (H-300) is recommended for detection of Dyskerin of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for Dyskerin siRNA (h): sc-38254, Dyskerin siRNA (m): sc-38255, Dyskerin shRNA Plasmid (h): sc-38254-SH, Dyskerin shRNA Plasmid (m): sc-38255-SH, Dyskerin shRNA (h) Lentiviral Particles: sc-38254-V and Dyskerin shRNA (m) Lentiviral Particles: sc-38255-V.

Dyskerin (H-300) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

Molecular Weight of Dyskerin: 58 kDa.

Positive Controls: HeLa nuclear extract: sc-2120 or MCF7 nuclear extract: sc-2149.

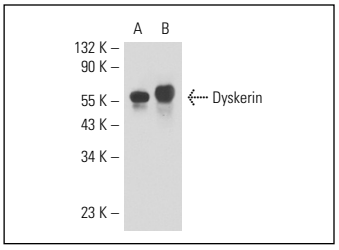
RESEARCH USE

For research use only, not for use in diagnostic procedures.

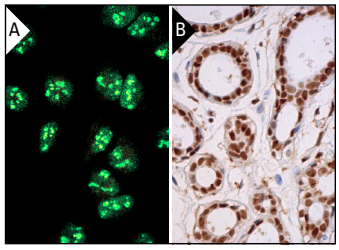
STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

DATA



Dyskerin (H-300): sc-48794. Western blot analysis of Dyskerin expression in HeLa (**A**) and MCF7 (**B**) nuclear extracts.



Dyskerin (H-300): sc-48794. Immunofluorescence staining of formalin-fixed HeLa cells showing nucleolar localization. Kindly provided by Dr. Nobuaki Kikyo, Stem Cell Institute, University of Minnesota (**A**). Immunoperoxidase staining of formalin fixed, paraffin-embedded human breast tissue showing nuclear staining of glandular cells (**B**).

SELECT PRODUCT CITATIONS

1. Alawi, F. and Lee, M.N. 2007. DKC1 is a direct and conserved transcriptional target of c-Myc. Biochem. Biophys. Res. Commun. 362: 893-898.
2. Trahan, C. and Dragon, F. 2009. Dyskeratosis congenita mutations in the H/ACA domain of human telomerase RNA affect its assembly into a pre-RNP. RNA 15: 235-243.
3. Parry, E.M., et al. 2011. Decreased dyskerin levels as a mechanism of telomere shortening in X-linked dyskeratosis congenita. J. Med. Genet. 48: 327-333.
4. Lattmann, S., et al. 2011. The DEAH-box RNA helicase RHAU binds an intramolecular RNA G-quadruplex in TERC and associates with telomerase holoenzyme. Nucleic Acids Res. 39: 9390-9404.
5. Gardano, L., et al. 2012. Native gel electrophoresis of human telomerase distinguishes active complexes with or without dyskerin. Nucleic Acids Res. 40: e36.
6. Alawi, F. and Lin, P. 2013. Dyskerin localizes to the mitotic apparatus and is required for orderly mitosis in human cells. PLoS ONE 8: e80805.
7. Schertzer, M., et al. 2015. Human regulator of telomere elongation helicase 1 (RTEL1) is required for the nuclear and cytoplasmic trafficking of pre-U2 RNA. Nucleic Acids Res. 43: 1834-1847.
8. Jeong, S.A., et al. 2015. Akt-mediated phosphorylation increases the binding affinity of hTERT for importin α to promote nuclear translocation. J. Cell Sci. 128: 2287-2301.