

Dyskerin (H-3): sc-373956

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-3) is a mouse monoclonal antibody raised against amino acids 171-470 mapping within an internal region of Dyskerin of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin. Also available as TransCruz reagent for Gel Supershift and ChIP applications, sc-373956 X, 200 µg/0.1 ml.

Dyskerin (H-3) is available conjugated to agarose (sc-373956 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-373956 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-373956 PE), fluorescein (sc-373956 FITC), Alexa Fluor® 488 (sc-373956 AF488), Alexa Fluor® 546 (sc-373956 AF546), Alexa Fluor® 594 (sc-373956 AF594) or Alexa Fluor® 647 (sc-373956 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-373956 AF680) or Alexa Fluor® 790 (sc-373956 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

Dyskerin (H-3) is recommended for detection of Dyskerin of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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) 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-3) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

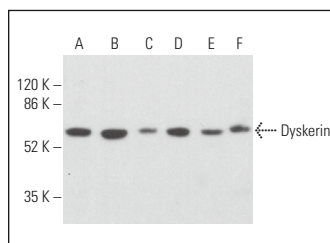
Molecular Weight of Dyskerin: 58 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, Raji whole cell lysate: sc-364236 or SW480 cell lysate: sc-2219.

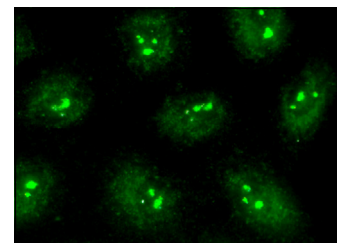
STORAGE

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

DATA



Dyskerin (H-3): sc-373956. Western blot analysis of Dyskerin expression in HeLa (A), Raji (B), Hep G2 (C), SW480 (D), HCT-116 (E) and MCF7 (F) whole cell lysates. Detection reagent used: m-IgGκ-BP-HRP: sc-516102.



Dyskerin (H-3): sc-373956. Immunofluorescence staining of methanol-fixed HeLa cells showing nucleolar localization.

SELECT PRODUCT CITATIONS

- Tang, M., et al. 2015. Disease mutant analysis identifies a new function of DAXX in telomerase regulation and telomere maintenance. *J. Cell Sci.* 128: 331-341.
- Poole, A.R. and Hebert, M.D. 2016. SMN and coilin negatively regulate Dyskerin association with telomerase RNA. *Biol. Open* 5: 726-735.
- Deng, T., et al. 2019. TOE1 acts as a 3' exonuclease for telomerase RNA and regulates telomere maintenance. *Nucleic Acids Res.* 47: 391-405.
- Patrick, E.M., et al. 2020. Observation of processive telomerase catalysis using high-resolution optical tweezers. *Nat. Chem. Biol.* 16: 801-809.
- García-Castillo, J., et al. 2021. Telomerase RNA recruits RNA polymerase II to target gene promoters to enhance myelopoiesis. *Proc. Natl. Acad. Sci. USA* 118: e2015528118.
- Zacchini, F., et al. 2022. Human dyskerin binds to cytoplasmic H/ACA-box-containing transcripts affecting nuclear hormone receptor dependence. *Genome Biol.* 23: 177.
- Sochacka, M., et al. 2022. FGF12 is a novel component of the nucleolar NOLC1/TCOF1 ribosome biogenesis complex. *Cell Commun. Signal.* 20: 182.
- Ranhem, C., et al. 2022. Evaluation of Dyskerin expression and the Cajal body protein WRAP53β as potential prognostic markers for patients with primary vaginal carcinoma. *Oncol. Lett.* 23: 30.
- Song, J., et al. 2023. CRISPR-free, programmable RNA pseudouridylation to suppress premature termination codons. *Mol. Cell* 83: 139-155.e9.
- Klump, B.M., et al. 2023. TCAB1 prevents nucleolar accumulation of the telomerase RNA to facilitate telomerase assembly. *Cell Rep.* 42: 112577.

RESEARCH USE

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

Alexa Fluor® is a trademark of Molecular Probes, Inc., Oregon, USA