SANTA CRUZ BIOTECHNOLOGY, INC.

Dyskerin (N-17): sc-26980

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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.

REFERENCES

1. Hassock, S., et al. 1999. Mapping and characterization of the X-linked dyskeratosis congenita (DKC) gene. Genomics 55: 21-27.
2. Heiss, N.S., et al. 1999. Dyskerin localizes to the nucleolus and its mislocalization is unlikely to play a role in the pathogenesis of dyskeratosis congenita. Hum. Mol. Genet. 8: 2515-2524.
3. Dragon, F., et al. 2000. *In vitro* assembly of human H/ACA small nucleolar RNPs reveals unique features of U17 and telomerase RNAs. Mol. Cell. Biol. 20: 3037-3048.
4. Pogacic, V., et al. 2000. Human H/ACA small nucleolar RNPs and telomerase share evolutionarily conserved proteins NHP2 and NOP10. Mol. Cell. Biol. 20: 9028-9040.
5. Wang, C., et al. 2002. Immunopurified small nucleolar ribonucleoprotein particles pseudouridylate rRNA independently of their association with phosphorylated Nopp140. Mol. Cell. Biol. 22: 8457-8466.
6. LocusLink Report (LocusID: 54433). http://www.ncbi.nlm.nih.gov/LocusLink/

CHROMOSOMAL LOCATION

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

SOURCE

Dyskerin (N-17) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus 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.

Blocking peptide available for competition studies, sc-26980 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

Dyskerin (N-17) 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), 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).

Dyskerin (N-17) is also recommended for detection of Dyskerin in additional species, including equine, canine, bovine and porcine.

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.

Molecular Weight of Dyskerin: 58 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.