

# Product datasheet for AP06643PU-N

# **DKC1 Rabbit Polyclonal Antibody**

**Product data:** 

**Product Type: Primary Antibodies** 

**Applications:** IF, IHC, WB

Recommended Dilution: Western blot: 1/500-1/1000.

Immunofluorescence: 1/50-1/200.

**Immunohistochemistry on Paraffin Sections:** 1/50-1/200.

Reactivity: Human, Mouse, Rat

Host: Rabbit

Clonality: Polyclonal

Immunogen: Synthetic peptide, corresponding to amino acids 171-220 of Human Dyskerin.

The antibody detects endogenous levels of Dyskerin protein. Specificity:

(region surrounding Lys203)

Formulation: Phosphate buffered saline (PBS), pH 7.2.

State: Aff - Purified

State: Liquid purified Ig fraction (> 95% by SDS-PAGE)

Preservative: 0.05% Sodium Azide

Concentration: 1.0 mg/ml

**Purification:** Affinity Chromatography using epitope-specific immunogen

Conjugation: Unconjugated

Storage: Store undiluted at 2-8°C for one month or (in aliquots) at -20°C for longer.

Avoid repeated freezing and thawing.

Stability: Shelf life: one year from despatch.

**Predicted Protein Size:** ~ 57 kDa

Gene Name: dyskerin pseudouridine synthase 1

Database Link: Entrez Gene 1736 Human

O60832



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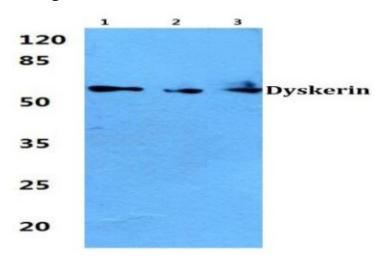
### Background:

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 and encodes a 28 kDa protein. The NHP2 gene maps to chromosome 5q35.3 and encodes a 155 amino acid protein.

Synonyms:

Dyskerin, Nucleolar protein NAP57, CBF5 homolog

## **Product images:**



Western blot (WB) analysis of Dyskerin antibody at 1/500 dilution Lane 1:A549 cell lysate Lane 2:Mouse brain tissue lysate Lane 3:Rat brain tissue lysate