

产品名称: Dyskerin Rabbit Polyclonal Antibody

产品货号: APRab10244

产品概述 (Summary)

描述(Description)	Rabbit polyclonal Antibody
宿主(Host)	Rabbit
应用(Application)	WB,IHC,ICC/IF,ELISA
种属反应(Reactivity)	Human,Mouse,Rat
偶联物(Conjugation)	Unconjugated
修饰(Modification)	Unmodified
同种型(Isotype)	IgG
克隆(Clonality)	Polyclonal
剂型(Form)	Liquid
保存条件(Storage)	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
储存溶液(Buffer)	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
纯化 (Purification)	Affinity purification

产品应用(Application)

稀释比(Dilution Ratio)	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:200-1:1000,ELISA 1:20000-1:40000
分子量(Molecular Weight)	57kDa

抗原信息(Antigen Information)

基因名(Gene Name)	DKC1
别名(Alternative Names)	DKC1; NOLA4; H/ACA ribonucleoprotein complex subunit 4; CBF5 homolog; Dyskerin; Nopp140-associated protein of 57 kDa; Nucleolar protein NAP57; Nucleolar protein family A member 4; snoRNP protein DKC1
基因 ID(Gene ID)	1736.0
SwissProt ID	O60832
免疫原(Immunogen)	The antiserum was produced against synthesized peptide derived from human Dyskerin. AA range:171-220

研究背景 (Background)

dyskerin pseudouridine synthase 1(DKC1) Homo sapiens This gene functions in two distinct complexes. It plays an active role in telomerase stabilization and maintenance, as well as recognition of snoRNAs containing H/ACA sequences which provides stability during biogenesis and assembly into H/ACA small nucleolar RNA ribonucleoproteins (snoRNPs). This gene is highly conserved and widely expressed, and may play additional roles in nucleo-cytoplasmic shuttling, DNA damage response, and cell adhesion. Mutations have been associated with X-linked dyskeratosis congenita. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Jan 2014],catalytic activity:RNA uridine = RNA pseudouridine.,disease:Defects in DKC1 are a cause of dyskeratosis congenita X-linked recessive (XDKC) [MIM:305000]. XDKC is a rare, progressive bone marrow failure syndrome characterized by the triad of reticulated skin hyperpigmentation, nail dystrophy, and mucosal leukoplakia. Early mortality is often associated with bone marrow failure, infections, fatal pulmonary complications, or malignancy.,disease:Defects in DKC1 are the cause of Hoyerall-Hreidarsson syndrome (HHS) [MIM:300240]. HHS is a multisystem disorder affecting males and is characterized by aplastic anemia, immunodeficiency, microcephaly, cerebellar hypoplasia, and growth retardation.,function:Required for ribosome biogenesis and telomere maintenance. Probable catalytic subunit of H/ACA small nucleolar ribonucleoprotein (H/ACA snoRNP) complex, which catalyzes pseudouridylation of rRNA. This involves the isomerization of uridine such that the ribose is subsequently attached to C5, instead of the normal N1. Each rRNA can contain up to 100 pseudouridine ('psi') residues, which may serve to stabilize the conformation of rRNAs. Also required for correct processing or intranuclear trafficking of TERC, the RNA component of the telomerase reverse transcriptase (TERT) holoenzyme.,online information:DKC1 mutation db,similarity:Belongs to the pseudouridine synthase truB family.,similarity:Contains 1 PUA domain.,subcellular location:Also localized to Cajal bodies (coiled bodies),subunit:Part of the H/ACA small nucleolar ribonucleoprotein (H/ACA snoRNP) complex, which contains NHP2/NOLA2, GAR1/NOLA1, NOP10/NOLA3, and DKC1/NOLA4, which is presumed to be the catalytic subunit. The complex contains a stable core formed by binding of one or two NOP10-DKC1 heterodimers to NHP2; GAR1 subsequently binds to this core via DKC1. The complex binds a box H/ACA small nucleolar RNA (snoRNA), which may target the specific site of modification within the RNA substrate. During assembly, the complex contains NAF1 instead of GAR1/NOLA1. The complex also interacts with TERC, which contains a 3'-terminal domain related to the box H/ACA snoRNAs. Specific interactions with snoRNAs or TERC are mediated by GAR1 and NHP2. Associates with NOLC1/NOPP140. H/ACA snoRNPs interact with the SMN complex, consisting of SMN1 or SMN2, SIP1/GEMIN2, DDX20/GEMIN3, and GEMIN4. This is mediated by interaction between GAR1 and SMN1 or SMN2. The SMN complex may be required for correct assembly of the H/ACA snoRNP complex. Component of the telomerase holoenzyme complex at least composed of TERT, DKC1, WDR79/TCAB1, NOP10, NHP2, GAR1, TEP1, EST1A, POT1 and a telomerase RNA template component (TERC),tissue specificity:Ubiquitously expressed.,

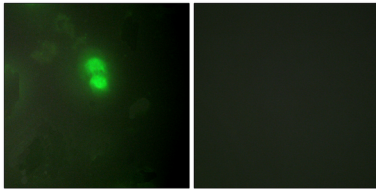
研究领域 (Research Area)

Epigenetics and Nuclear Signaling

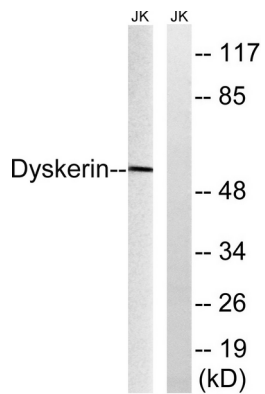
注意事项 (Note)

For research use only.

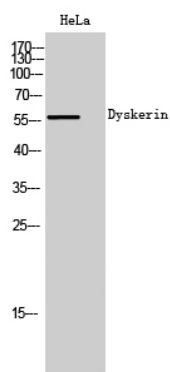
图片 (Image Data)



Immunofluorescence analysis of HeLa cells, using Dyskerin Antibody. The picture on the right is blocked with the synthesized peptide.



Western blot analysis of lysates from JurKat cells, using Dyskerin Antibody. The lane on the right is blocked with the synthesized peptide.



Western Blot analysis of HeLa cells using Dyskerin Polyclonal Antibody.

EnkiLife 优势产品推荐

产品类别	产品货号	产品名称
WB 解决方案	RA10020	2h 极速 WB 即用型全流程试剂盒
	RA10021	4h 快速 WB 即用型全流程试剂盒
	RA10042	5h 畅享版 WB 全流程试剂盒
	RA10037	校准级彩色预染蛋白 Marker (8-180kDa)
	RA10038	校准级彩色预染蛋白 Marker (10-250kDa)
	RA10039	校准级高分子彩色预染蛋白 Marker (25-400kDa)
TSA 多重荧光染色试剂盒	RA10008	TSA 双标三色多重荧光染色试剂盒 (mIHC)
	RA10009	TSA 三标四色多重荧光染色试剂盒 (mIHC)
	RA10010	TSA 四标五色多重荧光染色试剂盒 (mIHC)
	RA10011	TSA 五标六色多重荧光染色试剂盒 (mIHC)
	RA10012	TSA 六标七色多重荧光染色试剂盒 (mIHC)
IHC 检测试剂盒	RA10006	HRP Anti-Mouse/Rabbit IHC Detection System
	RA10007	Polymer-HRP Anti-Mouse/Rabbit IHC Detection System
抗体标记试剂盒	RE80004p	辣根过氧化物酶(HRP)抗体标记试剂盒
	RE80002q	Sulfo-NHS-生物素标记试剂盒
	RE80007p	Cy3 荧光素标记试剂盒
	RE80011p	Fluor488 荧光素标记试剂盒
	RE80017p	Fluor750 荧光素标记试剂盒
	RE80005p	藻红蛋白(R-PE) 抗体快速标记试剂盒
	RE80040	PE-Cy7 串联染料抗体快速标记试剂盒
稳转细胞系构建服务 (免费赠送全膜 WB 验证)	TS-0001	过表达稳转细胞系构建
	TS-0002	敲低稳转细胞系构建
	TS-0003	敲除细胞系构建

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