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ERCC8 Rabbit pAb

Catalog # AP99234

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	Q13216
Other Accession	Q13216
Reactivity	Rat, Mouse
Predicted	Human, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	44055
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human ERCC8/CSA
Epitope Specificity	161-260/396
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 5 WD repeats.
SUBUNIT	Part of the CSA complex (DCX(ERCC8) complex), a DCX E3 ubiquitin-protein ligase complex containing ERCC8, RBX1, DDB1 and CUL4A; the CSA complex interacts with RNA polymerase II; upon UV irradiation it interacts with the COP9 signalosome and preferentially with the hyperphosphorylated form of RNA polymerase II. Interacts with ERCC6 and KIAA1530/UVSSA. Interacts with a subunit of RNA polymerase II TFIIH. Interacts with DDB1.
DISEASE	Defects in ERCC8 are the cause of Cockayne syndrome type A (CSA) [MIM:216400]. Cockayne syndrome is a rare disorder characterized by cutaneous sensitivity to sunlight, abnormal and slow growth, cachectic dwarfism, progeroid appearance, progressive pigmentary retinopathy and sensorineural deafness. There is delayed neural development and severe progressive neurologic degeneration resulting in mental retardation. Two clinical forms are recognized: in the classical form or Cockayne syndrome type 1, the symptoms are progressive and typically become apparent within the first few years or life; the less common Cockayne syndrome type 2 is characterized by more severe symptoms that manifest prenatally. Cockayne syndrome shows some overlap with certain forms of xeroderma pigmentosum. Unlike xeroderma pigmentosum, patients with Cockayne syndrome do not manifest increased freckling and other pigmentation abnormalities in the skin and have no significant increase in skin cancer.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Nucleotide excision repair of DNA lesions occurs more rapidly and at a higher frequency on the template, or the transcribed, strand of DNA and to a much lesser extent on the coding, or the non-transcribed, strand or on transcriptionally inactive DNA. CSA and CSB are two related genes that are responsible for directing this preferential DNA repair pattern, known as transcriptional-repair coupling. Cells from patients with the UV-sensitive nucleotide excision repair disorder Cockayne's syndrome (CS) have specific mutations affecting these genes and results in defects of the preferential

repair on the transcribed strand of activated genes. CSA is a protein that belongs in the "WD-repeat" family of proteins. CSB, which is also designated excision repair cross-complementing protein-6 (ERCC-6), is the homolog of the yeast Rad26 protein. CSB belongs in the SWI/SNF family of proteins as it contains helicase motifs and ATPase activity.

Additional Information

Gene ID 1161

Other Names CKN1; CSA; UVSS2; 2410022P04Rik; 2810431L23Rik; 4631412O06Rik; B130065P18Rik; ERCC8_HUMAN; ERCC8; Cockayne syndrome WD repeat protein CSA; ERCC8_MOUSE; Cockayne syndrome WD repeat protein CSA homolog; ERCC excision repair 8, CSA ubiquitin ligase complex subunit; Cockayne syndrome 1 (classical); excision repair cross-complementing rodent repair deficiency, complementation group 8; excision repair cross-complementation group 8

Dilution WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500

Storage Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name ERCC8 {ECO:0000303|PubMed:19894250, ECO:0000312|HGNC:HGNC:3439}

Function Substrate-recognition component of the CSA complex, a DCX (DDB1-CUL4-X-box) E3 ubiquitin-protein ligase complex, involved in transcription-coupled nucleotide excision repair (TC-NER), a process during which RNA polymerase II-blocking lesions are rapidly removed from the transcribed strand of active genes (PubMed:12732143, PubMed:16751180, PubMed:16964240, PubMed:32142649, PubMed:34526721, PubMed:38316879, PubMed:38600235, PubMed:38600236). Following recruitment to lesion-stalled RNA polymerase II (Pol II), the CSA complex mediates ubiquitination of Pol II subunit POLR2A/RPB1 at 'Lys- 1268', a critical TC-NER checkpoint, governing RNA Pol II stability and initiating DNA damage excision by TFIIH recruitment (PubMed:12732143, PubMed:16751180, PubMed:16964240, PubMed:32142649, PubMed:32355176, PubMed:34526721, PubMed:38316879, PubMed:38600235, PubMed:38600236). The CSA complex also promotes the ubiquitination and subsequent proteasomal degradation of ERCC6/CSB in a UV-dependent manner; ERCC6 degradation is essential for the recovery of RNA synthesis after transcription-coupled repair (PubMed:16751180). Also plays a role in DNA double-strand breaks (DSSBs) repair by non-homologous end joining (NHEJ) (PubMed:29545921).

Cellular Location Nucleus. Chromosome Nucleus matrix. Note=Recruited to lesion- stalled RNA polymerase II (Pol II) sites by ERCC6/CSB (PubMed:32355176). UV-induced translocation to the nuclear matrix is dependent on ERCC6/CSB (PubMed:26620705).

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