ERCC8 Antibody – Rabbit Polyclonal

PRODUCT OVERVIEW

Product Name	ERCC8, Polyclonal Antibody
Full Product Name	ERCC8 (Center) Rabbit pAb
Product Gene Name	anti-ERCC8 antibody
Product Synonym Names	DNA excision repair protein ERCC-8; Cockayne syndrome WD repeat protein CSA; ERCC8; CKN1; CSA
Clonality	Polyclonal
Isotype	Ig
Clone Number	RB20950
Host	Rabbit
Species Reactivity	Human, Mouse
Form/Format	Supplied in PBS with 0.09% (W/V) Sodium Azide. This antibody is purified through a protein A column, followed by peptide affinity purification.
Purity/Purification	Affinity Purified
OMIM	gene 614621
3D Structure	ModBase 3D Structure for Q13216
Immunogen	This ERCC8 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 210-238 amino acids from the central region of human ERCC8.
Cellular Localization	Nucleus
ISO Certification	Manufactured in an ISO 9001:2015 Certified Laboratory.
Applications Tested/Suitable for anti-ERCC8 antibody	Western Blot (WB), Immunohistochemistry (IHC)
Application Notes for anti-ERCC8 antibody	WB: 1:1000 IHC: 1:50-100
Related Product Information for anti-ERCC8 antibody	MBS8551382 is an antibody targeting ERCC8. Also known as ERCC8 (Center) Rabbit pAb. Reactive in Human, Mouse. Host: Rabbit.
Other Notes	Small volumes of anti-ERCC8 antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for ERCC8. It may not necessarily be applicable to this product.

NCBI GI #	4557467
NCBI GeneID	1161
NCBI Accession #	NP_000073.1
UniProt Primary Accession #	Q13216
UniProt Related Accession #	Q13216

GenBank Nucleotide #	NP_000073.1
NCBI Official Full Name	DNA excision repair protein ERCC-8 isoform 1
NCBI Official Synonym Full Names	ERCC excision repair 8, CSA ubiquitin ligase complex subunit
NCBI Official Symbol	ERCC8
NCBI Official Synonym Symbols	CSA; CKN1; UVSS2
UniProt Protein Name	DNA excision repair protein ERCC-8
UniProt Synonym Protein Names	Cockayne syndrome WD repeat protein CSA
Protein Family	DNA excision repair protein
UniProt Gene Name	ERCC8
UniProt Synonym Gene Names	CKN1; CSA
UniProt Entry Name	ERCC8_HUMAN
NCBI Protein Information	DNA excision repair protein ERCC-8
NCBI Summary for ERCC8	This gene encodes a WD repeat protein, which interacts with Cockayne syndrome type B (CSB) protein and with p44 protein, a subunit of the RNA polymerase II transcription factor IIH. Mutations in this gene have been identified in patients with hereditary disease Cockayne syndrome (CS). CS cells are abnormally sensitive to ultraviolet radiation and are defective in the repair of transcriptionally active genes. Several transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, Mar 2014]
UniProt Comments for ERCC8	ERCC8: 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. The CSA complex (DCX(ERCC8) complex) promotes the ubiquitination and subsequent proteasomal degradation of ERCC6 in a UV-dependent manner; ERCC6 degradation is essential for the recovery of RNA synthesis after transcription-coupled repair. It is required for the recruitment of XAB2, HMGN1 and TCEA1/TFIIS to a transcription- coupled repair complex which removes RNA polymerase II-blocking lesions from the transcribed strand of active genes. Defects in ERCC8 are the cause of Cockayne syndrome type A (CSA). 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. Defects in ERCC8 are the cause of UV-sensitive syndrome type 2 (UVSS2). An autosomal recessive disorder characterized by cutaneous photosensitivity and mild freckling in the absence of neurological abnormalities or skin tumors. 2 isoforms of the human protein are produced by alternative splicing. Protein type: DNA repair, damage; Helicase; Ubiquitin conjugating system Chromosomal Location of Human Ortholog: 5q12.1 Cellular Component: nucleoplasm; nucleotide-excision repair complex; nuclear matrix; protein complex; nucleus Molecular Function: DNA-dependent ATPase activity; protein binding; DNA helicase activity; ubiquitin-protein ligase activity; protein complex binding Biological Process: proteasomal ubiquitin-dependent protein catabolic process; protein autoubiquitination; positive regulation of DNA repair; protein polyubiquitination; nucleotide-excision repair; transcription-coupled nucleotide-excision repair; response to oxidative stress; DNA repair; response to DNA damage stimulus; response to X-ray; response to UV Disease: Cockayne Syndrome A; Uv-sensitive Syndrome 2

Research Articles on ERCC8	**1.** 21 Han Chinese patients with Cockayne syndrome type A were investigated to identify mutations in ERCC8/ERCC6, of which thirteen cases with CS-A were identified with the mutations of ERCC8. 5 types mutations of ERCC8 were identified. Exon 4 rearrangement mutation and c.394_398delTTACA were the major mutations present in Han Chinese patients with CS-A, and also discovered three novel mutations.
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