abcam

Product datasheet

Recombinant Human ERCC8 protein ab152282

1 Image

Overview

Product name	Recombinant Human ERCC8 protein
Protein length	Full length protein

Description

Nature	Recombinant
Source	Wheat germ
Amino Acid Sequence	
Species	Human
Sequence	MLGFLSARQTGLEDPLRLRRAESTRRVLGLELNKDRD VERIHGGGINTLD IEPVEGRYMLSGGSDGVIVLYDLENSSRQSYYTCKAVC SIGRDHPDVHRY SVETVQWYPHDTGMFTSSSFDKTLKVWDTNTLQTAD VFNFEETVYSHHMS PVSTKHCLVAVGTRGPKVQLCDLKSGSCSHILQGIFILF QTATTLSKRFN KKKRY
Amino acids	1 to 205
Tags	GST tag N-Terminus

Specifications

Our Abpromise guarantee covers the use of **ab152282** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	ELISA
	Western blot
Form	Liquid
Additional notes	Protein concentration is above or equal to 0.05 mg/ml.

Preparation and Storage

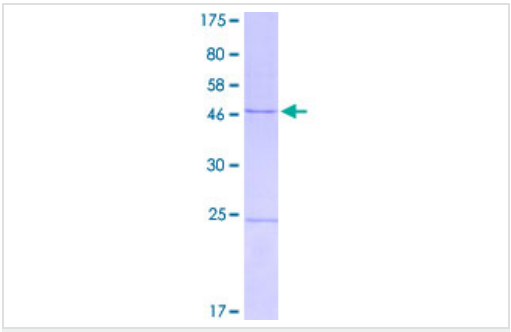
Stability and Storage	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles.

pH: 8.00
Constituents: 0.31% Glutathione, 0.79% Tris HCl

General Info

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. 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 recruitement 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.
Pathway	Protein modification; protein ubiquitination.
Involvement in 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.
Sequence similarities	Contains 5 WD repeats.
Cellular localization	Nucleus.

Images



SDS-PAGE - Recombinant Human ERCC8 protein (ab152282)

ab152282 on a 12.5% SDS-PAGE stained with Coomassie Blue.

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