abcam

Product datasheet

Anti-CSB antibody ab217202

1 Image

Overview

Product name	Anti-CSB antibody
Description	Rabbit polyclonal to CSB
Host species	Rabbit
Tested applications	**Suitable for:** IHC-P
Species reactivity	**Reacts with:** Rat **Predicted to work with:** Mouse, Human
Immunogen	Synthetic peptide within Human CSB aa 240-275 conjugated to keyhole limpet haemocyanin. The exact sequence is proprietary. Sequence: IRTGQMTPFGTQIPQKQEKKPRKIMLNEASGFEKYL Database link: Q03468 Run BLAST with Run BLAST with
Positive control	Rat kidney tissue.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term. Avoid freeze / thaw cycle.
Storage buffer	Preservative: 0.09% Sodium azide Constituents: 50% Glycerol, 1% BSA Aqueous buffered solution
Purity	Protein A purified
Clonality	Polyclonal
Isotype	IgG

Applications

Our Abpromise guarantee covers the use of **ab217202** in the following tested applications.

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

Application	Abreviews	Notes
IHC-P		1/100 - 1/500.

Target

Function

Essential factor involved in transcription-coupled nucleotide excision repair which allows RNA polymerase II-blocking lesions to be rapidly removed from the transcribed strand of active genes. Upon DNA-binding, it locally modifies DNA conformation by wrapping the DNA around itself, thereby modifying the interface between stalled RNA polymerase II and DNA. It is required for transcription-coupled repair complex formation. It recruits the CSA complex (DCX(ERCC8) complex), nucleotide excision repair proteins and EP300 to the at sites of RNA polymerase II-blocking lesions.

Involvement in disease

Defects in ERCC6 are the cause of Cockayne syndrome type B (CSB) [MIM:133540]. 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 ERCC6 are the cause of cerebro-oculo-facio-skeletal syndrome type 1 (COFS1) [MIM:214150]; also known as COFS syndrome or Pena-Shokeir syndrome type 2. COFS is a degenerative autosomal recessive disorder of prenatal onset affecting the brain, eye and spinal cord. After birth, it leads to brain atrophy, hypoplasia of the corpus callosum, hypotonia, cataracts, microcornea, optic atrophy, progressive joint contractures and growth failure. Facial dysmorphism is a constant feature. Abnormalities of the skull, eyes, limbs, heart and kidney also occur.
Defects in ERCC6 are a cause of De Sanctis-Cacchione syndrome (DSC) [MIM:278800]; also known as xerodermic idiocy. DSC is an autosomal recessive syndrome consisting of xeroderma pigmentosum associated with mental retardation, retarded growth, gonadal hypoplasia and sometimes neurologic complications.
Note=A genetic variation in the 5-prime flanking region of ERCC6 has been shown to be associated with susceptibility to age-related macular degeneration.
Defects in ERCC6 are a cause of UV-sensitive syndrome (UVS) [MIM:600630]. UVS is a rare autosomal recessive disorder characterized by photosensitivity and mild freckling but without neurological abnormalities or skin tumors.

Sequence similarities

Belongs to the SNF2/RAD54 helicase family.
Contains 1 helicase ATP-binding domain.
Contains 1 helicase C-terminal domain.

Domain

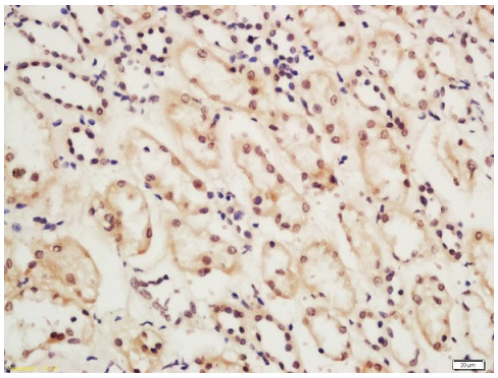
A C-terminal ubiquitin-binding domain (UBD) is essential for transcription-coupled nucleotide excision repair to proceed.

Post-translational modifications

Phosphorylated upon DNA damage, probably by ATM or ATR.
Ubiquitinated at the C-terminus. Ubiquitination by the CSA complex leads to ERCC6 proteasomal

	degradation in a UV-dependent manner.
Cellular localization	Nucleus.

Images



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-CSB antibody (ab217202)

Immunohistochemical analysis of formalin fixed, paraffin embedded Rat kidney tissue labeling CSB with ab217202 at 1/200 dilution followed by conjugation to the secondary antibody and DAB staining.

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