SunLong Biotech Co.,LTD
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[Primary Antibody]

Rabbit Anti-CSB antibody

Catalog Number: SL14082R

FOR RESEARCH USE ONLY

[DATASHEET]

Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	CSB
Immunogen:	KLH conjugated synthetic peptide derived from human CSB: 201-300/1493
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Applications:	IHC-P=1:100-500 IHC-F=1:100-500 IF=1:100-500 （Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Reactivity:	Rat(predicted:Human,Mouse)
Predicted MW.:	168kDa
Subcellular Location:	The nucleus
Background:	This gene encodes a DNA-binding protein that is important in transcription-coupled excision repair. The encoded protein has ATP-stimulated ATPase activity, interacts with several transcription and excision repair proteins, and may promote complex formation at DNA repair sites. Mutations in this gene are associated with Cockayne syndrome type B and cerebrooculofacioskeletal syndrome 1. Naturally-occurring readthrough transcription occurs between this gene and the adjacent PGBD3 gene (GeneID:267004), and results in a fusion protein that shares sequence with the product of each individual gene. The readthrough locus is represented by GeneID:101243544. [provided by RefSeq, Mar 2013] **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-

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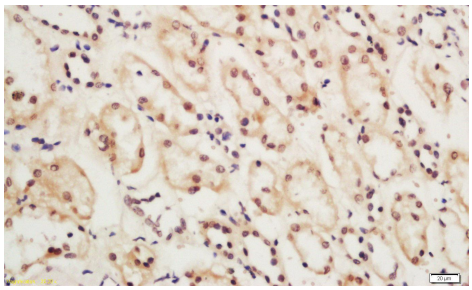
	binding, it locally modifies DNA conformation by wrapping the DNA around itself, thereby modifying the interface between stalled RNA polymerase II and DNA. It is required for transcription-coupled repair complex formation. It recruits the CSA complex (DCX(ERCC8) complex), nucleotide excision repair proteins and EP300 to the at sites of RNA polymerase II-blocking lesions. **Subcellular Location:** Nucleus. **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. 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)

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	[MIM:600630]. UVS is a rare autosomal recessive disorder characterized by photosensitivity and mild freckling but without neurological abnormalities or skin tumors. **Similarity:** Belongs to the SNF2/RAD54 helicase family. Contains 1 helicase ATP-binding domain. Contains 1 helicase C-terminal domain. **Database links:** **Entrez Gene:** 2074 Human **Entrez Gene:** 319955 Mouse Omim: 609413 Human **SwissProt:** Q03468 Human **SwissProt:** Q5W0L9 Human **SwissProt:** Q8C851 Mouse **Unigene:** 654449 Human

[VALIDATION IMAGES]



Tissue/cell: rat kidney tissue; 4% Paraformaldehyde-fixed and paraffin-embedded;
Antigen retrieval: citrate buffer (1M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min;
Incubation: Anti-CSB Polyclonal Antibody, Unconjugated(SL14082R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining

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