abcam

Product datasheet

Anti-CSB antibody ab96089

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製品の概要

製品名	Anti-CSB antibody
製品の詳細	Rabbit polyclonal to CSB
由来種	Rabbit
アプリケーション	**適用あり:** WB, ICC/IF
種交差性	**交差種:** Human **交差が予測される動物種:** Mouse
免疫原	Recombinant fragment within Human CSB aa 300-750. The exact immunogen sequence used to generate this antibody is proprietary information. If additional detail on the immunogen is needed to determine the suitability of the antibody for your needs, please **contact** our Scientific Support team to discuss your requirements.
ポジティブ・コントロール	A431 whole cell lysate and in culture. H1299 whole cell lysate. 293T, H1299, and HepG2.
特記事項	The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing. If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
バッファー	pH: 7.00 Preservative: 0.025% Proclin 300 Constituents: 50% Glycerol (glycerin, glycerine), 2.4% Tris, 1.5% Glycine, 46% PBS, 0.04% EGTA
精製度	Immunogen affinity purified
ポリ/モノ	ポリクローナル
アイソタイプ	IgG

アプリケーション

The Abpromise guarantee Abpromise保証は、次のテスト済みアプリケーションにおけるab96089の使用に適用されます

アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
WB	★★★★☆ (1)	1/500 - 1/3000. Predicted molecular weight: 168 kDa.
ICC/IF		1/100 - 1/200.

ターゲット情報

機能

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.

関連疾患

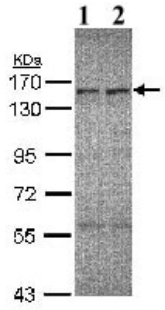
Defects in ERCC6 are the cause of Cockayne syndrome type B (CSB) [MIM:133540]. Cockayne syndrome is a rare disorder characterized by cutaneous sensitivity to sunlight, abnormal and slow growth, cachectic dwarfism, progeroid appearance, progressive pigmentary retinopathy and sensorineural deafness. There 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.

配列類似性

Belongs to the SNF2/RAD54 helicase family.

	Contains 1 helicase ATP-binding domain. Contains 1 helicase C-terminal domain.
ドメイン	A C-terminal ubiquitin-binding domain (UBD) is essential for transcription-coupled nucleotide excision repair to proceed.
翻訳後修飾	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.
細胞内局在	Nucleus.

画像



Western blot - Anti-CSB antibody (ab96089)

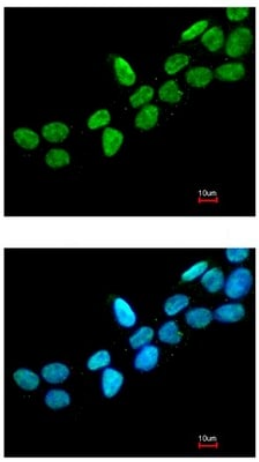
All lanes : Anti-CSB antibody (ab96089) at 1/500 dilution

Lane 1 : A431 whole cell lysate
Lane 2 : H1299 whole cell lysate

Lysates/proteins at 30 µg per lane.

Predicted band size: 168 kDa

7.5% SDS Page



Immunocytochemistry/ Immunofluorescence - Anti-CSB antibody (ab96089)

Immunofluorescence analysis of paraformaldehyde-fixed A431, using ab96089 antibody at 1/200 dilution. Lower image merged with DNA probe.

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