abcam

Product datasheet

Anti-CSB antibody ab87835

1 Abreviews 1 图像

概述

产品名称	Anti-CSB抗体
描述	**兔多克隆抗体**to CSB
宿主	Rabbit
经测试应用	**适用于:** WB
种属反应性	**与反应:** Human **预测可用于:** Chicken, Cow, Dog
免疫原	Synthetic peptide. This information is proprietary to Abcam and/or its suppliers.
阳性对照	This antibody gave a positive signal in the following lysates: HeLa Whole Cell Lysate - Irradiated (5Gy).
常规说明	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

性能

形式	Liquid
存放说明	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.
存储溶液	pH: 7.40 Preservative: 0.02% Sodium azide Constituent: PBS Batches of this product that have a concentration < 1mg/ml may have BSA added as a stabilising agent. If you would like information about the formulation of a specific lot, please contact our scientific support team who will be happy to help.
纯度	Immunogen affinity purified
克隆	**多克隆**

同种型	IgG

应用

The Abpromise guarantee **Abpromise™承诺保证使用ab87835于以下的经测试应用**

"应用说明"部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
WB		Use a concentration of 1 µg/ml. Detects a band of approximately 175 kDa (predicted molecular weight: 168 kDa).

靶标

功能

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.

疾病相关

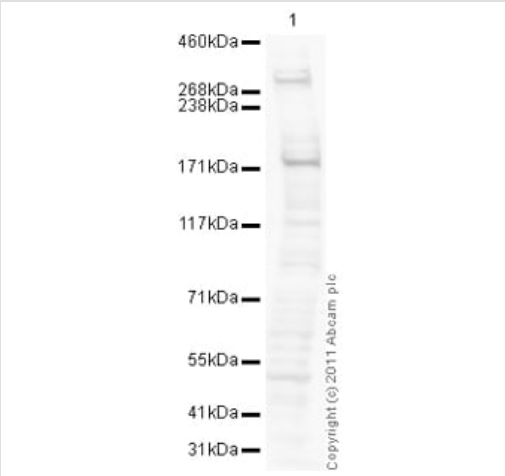
Defects in ERCC6 are the cause of Cockayne syndrome type B (CSB) [MIM:133540]. Cockayne syndrome is a rare disorder characterized by cutaneous sensitivity to sunlight, abnormal and slow growth, cachectic dwarfism, progeroid appearance, progressive pigmentary retinopathy and sensorineural deafness. There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

序列相似性

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

	Contains 1 helicase C-terminal domain.
结构域	A C-terminal ubiquitin-binding domain (UBD) is essential for transcription-coupled nucleotide excision repair to proceed.
翻译后修饰	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.
细胞定位	Nucleus.

图片



Western blot - Anti-CSB antibody (ab87835)

Anti-CSB antibody (ab87835) at 1 µg/ml + HeLa Whole Cell Lysate - Irradiated (5Gy) at 10 µg

Secondary
Goat Anti-Rabbit IgG H&L (HRP) preadsorbed (**ab97080**) at 1/5000 dilution

Developed using the ECL technique.

Performed under reducing conditions.

Predicted band size: 168 kDa
Observed band size: 175 kDa
Additional bands at: 300 kDa, 50 kDa. We are unsure as to the identity of these extra bands.

Exposure time: 150 seconds

Abcam recommends loading unheated lysates and using 20% Ethanol and 0.04% SDS in the transfer buffer to obtain a positive signal for this antibody. Abcam welcomes customer feedback and would appreciate any comments regarding this product and the data presented above

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