abcam

Product datasheet

Anti-CSB antibody [553C5a] ab66598

4 References 2 Images

Overview

Product name	Anti-CSB antibody [553C5a]
Description	Mouse monoclonal [553C5a] to CSB
Host species	Mouse
Tested applications	**Suitable for:** Flow Cyt, WB
Species reactivity	**Reacts with:** Human, Recombinant fragment
Immunogen	Recombinant fragment corresponding to Human CSB (internal sequence). Carrying 50-200 aa
General notes	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

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
Storage buffer	pH: 7.40 Preservative: 0.05% Sodium azide Constituents: PBS, 0.0225% Potassium chloride, 0.03% Potassium phosphate, 0.1312% Sodium phosphate, 0.812% Sodium chloride, 1% BSA
Purity	Protein G purified
Purification notes	Purified from culture supernatant of hybridoma cultured in a medium containing bovine IgG-depleted (approximately 95%) fetal bovine serum and filtered through a 0.22 µm membrane.
Clonality	Monoclonal
Clone number	553C5a
Isotype	IgG2b

Applications

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab66598 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
Flow Cyt		Use 1µg for 10^6 cells. **ab170192** - Mouse monoclonal IgG2b, is suitable for use as an isotype control with this antibody.
WB		1/100. Predicted molecular weight: 168 kDa.

Target

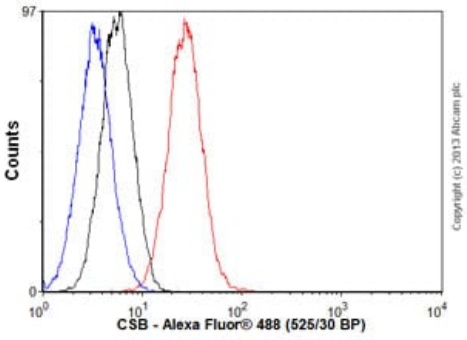
Function Essential factor involved in transcription-coupled nucleotide excision repair which allows RNA polymerase II-blocking lesions to be rapidly removed from the transcribed strand of active genes. Upon DNA-binding, it locally modifies DNA conformation by wrapping the DNA around itself, thereby modifying 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.

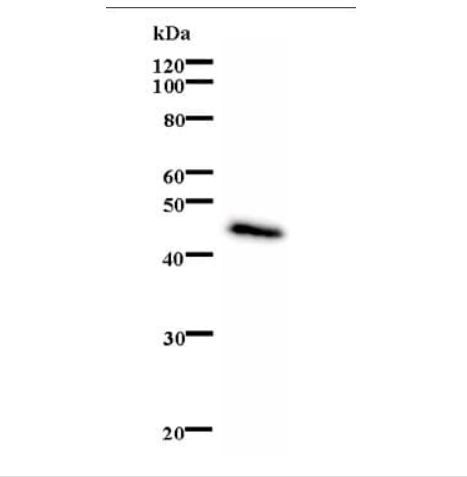
	Contains 1 helicase C-terminal domain.
Domain	A C-terminal ubiquitin-binding domain (UBD) is essential for transcription-coupled nucleotide excision repair to proceed.
Post-translational modifications	Phosphorylated upon DNA damage, probably by ATM or ATR. Ubiquitinated at the C-terminus. Ubiquitination by the CSA complex leads to ERCC6 proteasomal degradation in a UV-dependent manner.
Cellular localization	Nucleus.

Images



Flow Cytometry - Anti-CSB antibody [553C5a] (ab66598)

Overlay histogram showing HeLa cells stained with ab66598 (red line). The cells were fixed with 80% methanol (5 min) and then permeabilized with 0.1% PBS-Tween for 20 min. The cells were then incubated in 1x PBS / 10% normal goat serum / 0.3M glycine to block non-specific protein-protein interactions followed by the antibody (ab66598, 1µg/1x10^6 cells) for 30 min at 22°C. The secondary antibody used was Alexa Fluor® 488 goat anti-mouse IgG (H&L) (**ab150113**) at 1/2000 dilution for 30 min at 22°C. Isotype control antibody (black line) was mouse IgG2b [PLPV219] (**ab91366**, 1µg/1x10^6 cells) used under the same conditions. Unlabelled sample (blue line) was also used as a control. Acquisition of >5,000 events were collected using a 20mW Argon ion laser (488nm) and 525/30 bandpass filter.



Western blot - Anti-CSB antibody [553C5a] (ab66598)

Anti-CSB antibody [553C5a] (ab66598) + immunising recombinant protein

Predicted band size: 168 kDa
Observed band size: 45 kDa

Primary antibody dilution 1:100, Recombinant protein amount 10ng/lane, Secondary antibody dilution 1:3000, Secondary antibody Sheep anti mouse IgG (0.63mg/ml)

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