abcam

Product datasheet

Recombinant Human IKK gamma/NEMO protein ab206008

1 Image

Description

Product name	Recombinant Human IKK gamma/NEMO protein
Purity	>= 85 % SDS-PAGE.
Expression system	Escherichia coli
Accession	Q9Y6K9-1
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MNRHLWKSQLCEMVQPSGGPAADQDVLGEESPLGKPA MLHLPSEQGAPET LQRCLEENQELRDAIRQSNQILRERCEELLHFQASQREEK EFLMCKFQEA RKLVERLGLEKLDLKRQKEQALREVEHLKRCQQQMAED KASVKAQVTSLL GELQESQSRLEAATKECQALEGRARAASEQ ARQLESEREALQQQHSVQ VDQLRMQGQSVEAALRMERQAASEEKRKLAQLQVAYHQ LFQEYDNHIKSS VVGSERKRGMQLEDLKQQLQQAEEALVAKQEVIDKLKEE AEQHKIVMETV PVLKAQADIYKADFQAERQAREKLAEKKELLQEQLEQLQ REYSKLKASCQ ESARIEDMRKRHVEVSQAPLPPAPAYLSSPLALPSQRRS PPEEPPDFCCP KCQYQAPDMDTLQIHVMECIE
Predicted molecular weight	48 kDa
Amino acids	1 to 419
Tags	GST tag N-Terminus
Additional sequence information	NM_001099857

Specifications

Our Abpromise guarantee covers the use of **ab206008** in the following tested applications.

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

Applications	SDS-PAGE
Form	Liquid

Preparation and Storage

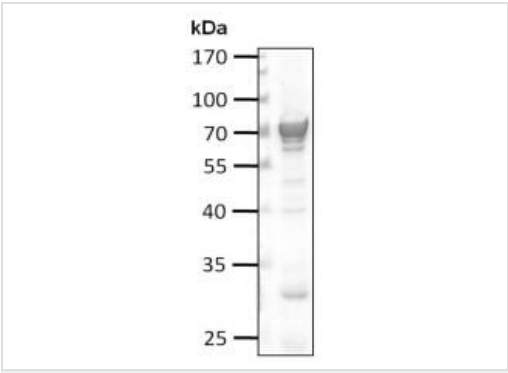
Stability and Storage	Shipped on Dry Ice. Store at -80°C. Avoid freeze / thaw cycle. Constituents: 0.32% Tris, 0.87% Sodium chloride, 0.15% Beta mercaptoethanol, 10% Glycerol

General Info

Function	Regulatory subunit of the IKK core complex which phosphorylates inhibitors of NF-kappa-B thus leading to the dissociation of the inhibitor/NF-kappa-B complex and ultimately the degradation of the inhibitor. Also considered to be a mediator for TAX activation of NF-kappa-B. Could be implicated in NF-kappa-B-mediated protection from cytokine toxicity (By similarity). Essential for viral activation of IRF3.
Tissue specificity	Heart, brain, placenta, lung, liver, skeletal muscle, kidney and pancreas.
Involvement in disease	Defects in IKBKG are the cause of ectodermal dysplasia anhidrotic with immunodeficiency X-linked (EDAID) [MIM:300291]; also known as hypohidrotic ectodermal dysplasia with immunodeficiency (HED-ID). Is a form of ectoderma dysplasia, a heterogeneous group of disorders due to abnormal development of two or more ectodermal structures. Characterized by absence of sweat glands, sparse scalp hair, rare conical teeth and immunological abnormalities resulting in severe infectious diseases. Defects in IKBKG are the cause of ectodermal dysplasia anhidrotic with immunodeficiency-osteopetrosis-lymphedema (OLEDAID) [MIM:300301]. Defects in IKBKG are a cause of immunodeficiency NEMO-related without anhidrotic ectodermal dysplasia (NEMOID) [MIM:300584]; also called immunodeficiency without anhidrotic ectodermal dysplasia, isolated immunodeficiency or pure immunodeficiency. Patients manifest immunodeficiency not associated with other abnormalities, and resulting in increased infection susceptibility. Patients suffer from multiple episodes of infectious diseases. Defects in IKBKG are the cause of susceptibility to X-linked familial atypical micobacteriosis type 1 (AMCBX1) [MIM:300636]; also known as X-linked disseminated atypical mycobacterial infection type 1 or X-linked susceptibility to mycobacterial disease type 1. AMCBX1 is the X-linked recessive form of mendelian susceptibility to mycobacterial disease (MSMD). MSMD is a congenital syndrome resulting in predisposition to clinical disease caused by weakly virulent mycobacterial species, such as bacillus Calmette-Guerin vaccines and non-tuberculous, environmental mycobacteria. Patients are also susceptible to the more virulent species Mycobacterium tuberculosis. Defects in IKBKG are the cause of recurrent isolated invasive pneumococcal disease type 2 (IPD2) [MIM:300640]. Recurrent invasive pneumococcal disease (IPD) is defined as two episodes of IPD occurring at least 1 month apart, whether caused by the same or different serotypes or strains. Recurrent IPD occurs in at least 2% of patients in most series, making IPD the most important known risk factor for subsequent IPD. Defects in IKBKG are the cause of incontinentia pigmenti (IP) [MIM:308300]; formerly designed familial incontinentia pigmenti type II (IP2). IP is a genodermatosis usually prenatally lethal in males. In affected females, it causes abnormalities of the skin, hair, eyes, nails, teeth, skeleton,

	heart, and central nervous system. The prominent skin signs occur in four classic cutaneous stages: perinatal inflammatory vesicles, verrucous patches, a distinctive pattern of hyperpigmentation and dermal scarring.
Sequence similarities	Contains 1 C2HC-type zinc finger.
Domain	The leucine-zipper domain and the C2HC-type zinc-finger are essential for polyubiquitin binding and for the activation of IRF3.
Post-translational modifications	Phosphorylation at Ser-68 attenuates aminoterminal homodimerization. Polyubiquitinated on Lys-285 through 'Lys-63'; the ubiquitination is mediated by NOD2 and RIPK2 and probably plays a role in signaling by facilitating interactions with ubiquitin domain-containing proteins and activates the NF-kappa-B pathway. Polyubiquitinated on Lys-399 through 'Lys-63'; the ubiquitination is mediated by BCL10, MALT1 and TRAF6 and probably plays a role in signaling by facilitating interactions with ubiquitin domain-containing proteins and activates the NF-kappa-B pathway. Monoubiquitinated on Lys-277 and Lys-309; promotes nuclear export. Linear polyubiquitinated on Lys-285; the head-to-tail polyubiquitination is mediated by the LUBAC complex. Linear polyubiquitinated on Lys-309; the head-to-tail polyubiquitination is mediated by the LUBAC complex. Sumoylated on Lys-277 and Lys-309 by SUMO1; the modification results in phosphorylation of Ser-85 by ATM leading to a replacement of the sumoylation by mono-ubiquitination on these residues.
Cellular localization	Cytoplasm. Nucleus. Sumoylated NEMO accumulates in the nucleus in response to genotoxic stress.

Images



SDS-PAGE - Recombinant Human IKK gamma/NEMO protein (ab206008)

SDS-PAGE analysis of ab206008 (5µg) Coomassie-stained.

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