abcam

Product datasheet

Recombinant Human TGF beta Receptor II protein (Fc Chimera) ab223104

1 Image

Description

Product name Recombinant Human TGF beta Receptor II protein (Fc Chimera)

Purity > 95 % SDS-PAGE.

Endotoxin level < 1.000 Eu/μg
Expression system HEK 293 cells

Accession <u>P37173</u>

Protein length Protein fragment

Animal free No

Nature Recombinant

Species Human

Sequence TIPPHVQKSVNNDMIVTDNNGAVKFPQLCKFCDVRFSTCD

NQKSCMSNCS

ITSICEKPQEVCVAVWRKNDENITLETVCHDPKLPYHDFIL

EDAASPKCI

MKEKKKPGETFFMCSCSSDECNDNIIFSEEYNTSNPD

Predicted molecular weight 42 kDa including tags

Amino acids 23 to 159

Additional sequence information Fused with human IgG1 Fc tag at the C-terminus (NP_003233).

Specifications

Our Abpromise quarantee covers the use of ab223104 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 Lyophilized

Preparation and Storage

Stability and Storage Shipped at 4°C. Store at -20°C or -80°C. Avoid freeze / thaw cycle.

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pH: 7.4

Constituents: 0.75% Glycine, 5% Trehalose, 0.61% Tris, Sodium chloride, L-Arginine

Lyophilized from 0.22 µm filtered solution.

Reconstitution

Reconstitute with sterile deionized water to a concentration of 400 µg/ml.

General Info

Function

Transmembrane serine/threonine kinase forming with the TGF-beta type I serine/threonine kinase receptor, TGFBR1, the non-promiscuous receptor for the TGF-beta cytokines TGFB1, TGFB2 and TGFB3. Transduces the TGFB1, TGFB2 and TGFB3 signal from the cell surface to the cytoplasm and is thus regulating a plethora of physiological and pathological processes including cell cycle arrest in epithelial and hematopoietic cells, control of mesenchymal cell proliferation and differentiation, wound healing, extracellular matrix production, immunosuppression and carcinogenesis. The formation of the receptor complex composed of 2 TGFBR1 and 2 TGFBR2 molecules symmetrically bound to the cytokine dimer results in the phosphorylation and the activation of TGFRB1 by the constitutively active TGFBR2. Activated TGFBR1 phosphorylates SMAD2 which dissociates from the receptor and interacts with SMAD4. The SMAD2-SMAD4 complex is subsequently translocated to the nucleus where it modulates the transcription of the TGF-beta-regulated genes. This constitutes the canonical SMAD-dependent TGF-beta signaling cascade. Also involved in non-canonical, SMAD-independent TGF-beta signaling pathways.

Involvement in disease

Defects in TGFBR2 are the cause of hereditary non-polyposis colorectal cancer type 6 (HNPCC6) [MIM:614331]. Mutations in more than one gene locus can be involved alone or in combination in the production of the HNPCC phenotype (also called Lynch syndrome). Most families with clinically recognized HNPCC have mutations in either MLH1 or MSH2 genes. HNPCC is an autosomal, dominantly inherited disease associated with marked increase in cancer susceptibility. It is characterized by a familial predisposition to early onset colorectal carcinoma (CRC) and extra-colonic cancers of the gastrointestinal, urological and female reproductive tracts. HNPCC is reported to be the most common form of inherited colorectal cancer in the Western world, and accounts for 15% of all colon cancers. Cancers in HNPCC originate within benign neoplastic polyps termed adenomas. Clinically, HNPCC is often divided into two subgroups. Type I: hereditary predisposition to colorectal cancer, a young age of onset, and carcinoma observed in the proximal colon. Type II: patients have an increased risk for cancers in certain tissues such as the uterus, ovary, breast, stomach, small intestine, skin, and larynx in addition to the colon. Diagnosis of classical HNPCC is based on the Amsterdam criteria: 3 or more relatives affected by colorectal cancer, one a first degree relative of the other two; 2 or more generation affected; 1 or more colorectal cancers presenting before 50 years of age; exclusion of hereditary polyposis syndromes. The term "suspected HNPCC" or "incomplete HNPCC" can be used to describe families who do not or only partially fulfill the Amsterdam criteria, but in whom a genetic basis for colon cancer is strongly suspected. HNPCC6 is a type of colorectal cancer complying with the clinical criteria of HNPCC, except that the onset of cancer was beyond 50 years of age in all cases.

Defects in TGFBR2 are a cause of esophageal cancer (ESCR) [MIM:133239].

Defects in TGFBR2 are the cause of Loeys-Dietz syndrome type 1B (LDS1B) [MIM:610168].

LDS1 is an aortic aneurysm syndrome with widespread systemic involvement. The disorder is characterized by arterial tortuosity and aneurysms, craniosynostosis, hypertelorism, and bifid uvula or cleft palate. Other findings include exotropy, micrognathia and retrognathia, structural brain abnormalities, intellectual deficit, congenital heart disease, translucent skin, joint hyperlaxity and aneurysm with dissection throughout the arterial tree.

Defects in TGFBR2 are the cause of Loeys-Dietz syndrome type 2B (LDS2B) [MIM:610380]. An

aortic aneurysm syndrome with widespread systemic involvement. Physical findings include prominent joint laxity, easy bruising, wide and atrophic scars, velvety and translucent skin with easily visible veins, spontaneous rupture of the spleen or bowel, diffuse arterial aneurysms and dissections, and catastrophic complications of pregnancy, including rupture of the gravid uterus and the arteries, either during pregnancy or in the immediate postpartum period. LDS2 is characterized by the absence of craniofacial abnormalities with the exception of bifid uvula that can be present in some patients. Note=TGFBR2 mutations Cys-460 and His-460 have been reported to be associated with thoracic aortic aneurysms and dissection (TAAD). This phenotype, also known as thoracic aortic aneurysms type 3 (AAT3), is distinguised from LDS2B by having aneurysms restricted to thoracic aorta. As individuals carrying these mutations also exhibit descending aortic disease and aneurysms of other arteries (PubMed:16027248), they have been considered as LDS2B by the OMIM resource.

Sequence similarities

Belongs to the protein kinase superfamily. TKL Ser/Thr protein kinase family. TGFB receptor

subfamily.

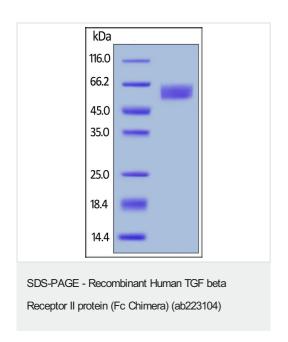
Contains 1 protein kinase domain.

Post-translational modifications

Phosphorylated on a Ser/Thr residue in the cytoplasmic domain.

Cellular localization Cell membrane.

Images



ab223104 analyzed by DTT-reduced SDS-PAGE and stained overnight with Coomassie Blue.

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