

Product datasheet

Recombinant human BTK (mutated C481R) protein (Active) ab268372

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Description

Product name	Recombinant human BTK (mutated C481R) protein (Active)
Biological activity	The specific activity of ab268372 was 2.5 nmol/min/mg in a kinase assay using Poly (4:1 Glu, Tyr) synthetic peptide as substrate.
Purity	> 75 % SDS-PAGE. Affinity purified.
Expression system	Baculovirus infected Sf9 cells
Accession	<u>Q06187</u>
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Molecular weight information	SDS-PAGE molecular weight: ~75kDa
Modifications	mutated C481R
Tags	His tag N-Terminus
Additional sequence information	GenBank: NM_000061

Specifications

Our **Abpromise guarantee** covers the use of **ab268372** in the following tested applications.

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

Applications	Functional Studies SDS-PAGE
Form	Liquid

Preparation and Storage

Stability and Storage	Shipped on Dry Ice. Upon delivery aliquot. Store at -80°C. Avoid freeze / thaw cycle. pH: 7.00 Preservative: 1.02% Imidazole
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Constituents: 0.82% Sodium phosphate, 1.74% Sodium chloride, 0.002% PMSF, 0.004% DTT, 25% Glycerol (glycerin, glycerine)

This product is an active protein and may elicit a biological response in vivo, handle with caution.

General Info

Function

Plays a crucial role in B-cell ontogeny. Transiently phosphorylates GTF2I on tyrosine residues in response to B-cell receptor cross-linking. Required for the formation of functional ARID3A DNA-binding complexes.

Involvement in disease

Defects in BTK are the cause of X-linked agammaglobulinemia (XLA) [MIM:300755]; also known as X-linked agammaglobulinemia type 1 (AGMX1) or immunodeficiency type 1 (IMD1). XLA is a humoral immunodeficiency disease which results in developmental defects in the maturation pathway of B-cells. Affected boys have normal levels of pre-B-cells in their bone marrow but virtually no circulating mature B-lymphocytes. This results in a lack of immunoglobulins of all classes and leads to recurrent bacterial infections like otitis, conjunctivitis, dermatitis, sinusitis in the first few years of life, or even some patients present overwhelming sepsis or meningitis, resulting in death in a few hours. Treatment in most cases is by infusion of intravenous immunoglobulin.

Defects in BTK may be the cause of X-linked hypogammaglobulinemia and isolated growth hormone deficiency (XLA-IGHD) [MIM:307200]; also known as agammaglobulinemia and isolated growth hormone deficiency or Fleisher syndrome or isolated growth hormone deficiency type 3 (IGHD3). In rare cases XLA is inherited together with isolated growth hormone deficiency (IGHD).

Sequence similarities

Belongs to the protein kinase superfamily. Tyr protein kinase family. TEC subfamily.

Contains 1 Btk-type zinc finger.

Contains 1 PH domain.

Contains 1 protein kinase domain.

Contains 1 SH2 domain.

Contains 1 SH3 domain.

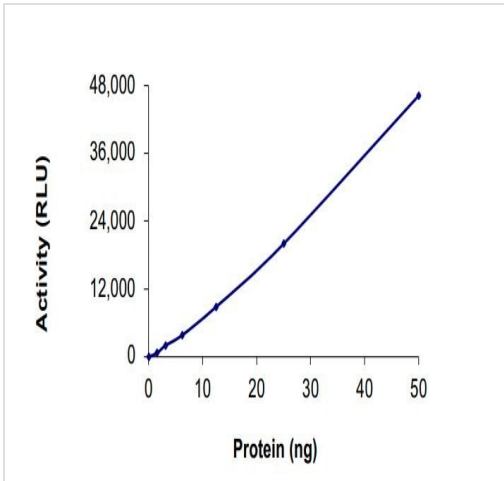
Post-translational modifications

Autophosphorylated on Tyr-223 and Tyr-551. Phosphorylation of Tyr-223 may create a docking site for a SH2 containing protein.

Cellular localization

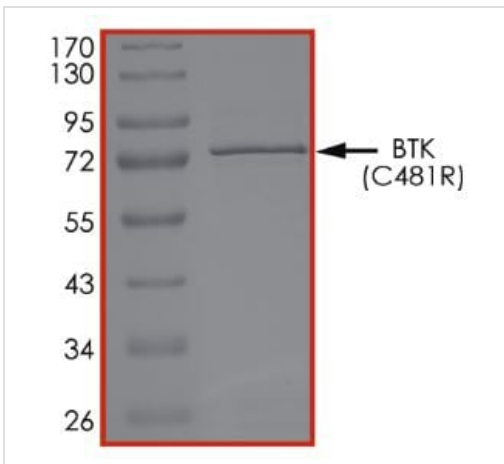
Cytoplasm. Membrane. Nucleus.

Images



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Functional Studies - Recombinant human BTK (mutated C481R) protein (Active) (ab268372)



SDS-PAGE analysis of ab268372.

SDS-PAGE - Recombinant human BTK (mutated C481R) protein (Active) (ab268372)

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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