

Product name:	Btk Rabbit Polyclonal Antibody
Cat number:	ABN07690
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	Synthesized peptide derived from Btk . at AA range: 490-570
Reactivity:	Human, Mouse, Rat
Applications:	WB 1:500-1:2000, IHC 1:100-1:300, ICC/IF 1:50-1:200, ELISA 1:10000-1:20000
Molecular Weight:	76kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

The protein encoded by this gene plays a crucial role in B-cell development. Mutations in this gene cause X-linked agammaglobulinemia type 1, which is an immunodeficiency characterized by the failure to produce mature B lymphocytes, and associated with a failure of Ig heavy chain rearrangement. Alternative splicing results in multiple transcript variants encoding different isoforms. [provided by RefSeq, Dec 2013], catalytic activity: ATP + a [protein]-L-tyrosine = ADP + a [protein]-L-tyrosine phosphate., cofactor: Binds 1 zinc ion per subunit., disease: Defects in BTK are the cause of X-linked agammaglobulinemia (XLA) [MIM:300755]; also called 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., disease: 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)., enzyme regulation: Inhibited by IBTK. Activated by phosphorylation., 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., online information: BTK mutation db, PTM: Autophosphorylated on Tyr-223 and Tyr-551. Phosphorylation of Tyr-223 may create a docking site for a SH2 containing protein., similarity: Belongs to the protein kinase superfamily. Tyr protein kinase family., similarity: Belongs to the protein kinase superfamily. Tyr protein kinase family. TEC subfamily., similarity: Contains 1 Btk-type zinc finger., similarity: Contains 1 PH domain., similarity: Contains 1 protein kinase domain., similarity: Contains 1 SH2 domain., similarity: Contains 1 SH3 domain., subunit: Binds GTF2I through the PH domain. Interacts with SH3BP5 via the SH3 domain. Interacts with IBTK via its PH domain. Interacts with GTF2I and ARID3A.,