

Product name:	BMPR-IB Rabbit Polyclonal Antibody
Cat number:	ABN07608
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	Synthetic peptide from human protein at AA range: 21-70
Reactivity:	Human, Mouse
Applications:	IHC 1:50-1:200, ICC/IF 1:50-1:200, ELISA 1:10000-1:20000
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:

This gene encodes a member of the bone morphogenetic protein (BMP) receptor family of transmembrane serine/threonine kinases. The ligands of this receptor are BMPs, which are members of the TGF-beta superfamily. BMPs are involved in endochondral bone formation and embryogenesis. These proteins transduce their signals through the formation of heteromeric complexes of 2 different types of serine (threonine) kinase receptors: type I receptors of about 50-55 kD and type II receptors of about 70-80 kD. Type II receptors bind ligands in the absence of type I receptors, but they require their respective type I receptors for signaling, whereas type I receptors require their respective type II receptors for ligand binding. Mutations in this gene have been associated with primary pulmonary hypertension. Several transcript variants encoding two different isoforms have been found for thicatalytic activity:ATP + [receptor-protein] = ADP + [receptor-protein] phosphate.,cofactor:Magnesium or manganese.,disease:Defects in BMPR1B are a cause of brachydactyly type A2 (BDA2) [MIM:112600]. Brachydactylies (BDs) are a group of inherited malformations characterized by shortening of the digits due to abnormal development of the phalanges and/or the metacarpals. They have been classified on an anatomic and genetic basis into five groups, A to E, including three subgroups (A1 to A3) that usually manifest as autosomal dominant traits. BDA2 was described first in a large Norwegian kindred. BDA2 is caused by mutations in BMPR1B gene and studies demonstrate that these mutations function as dominant negatives in vitro and in vivo.,disease:Defects in BMPR1B are the cause of acromesomelic chondrodysplasia with genital anomalies (AMDGA) [MIM:609441]. Acromesomelic chondrodysplasias are rare hereditary skeletal disorders characterized by short stature, very short limbs, and hand/foot malformations. The severity of limb abnormalities increases from proximal to distal with profoundly affected hands and feet showing brachydactyly and/or rudimentary fingers (knob-like fingers).,function:On ligand binding, forms a receptor complex consisting of two type II and two type I transmembrane serine/threonine kinases. Type II receptors phosphorylate and activate type I receptors which autophosphorylate, then bind and activate SMAD transcriptional regulators. Receptor for BMPS/OP-1.,similarity:Belongs to the protein kinase superfamily. TKL Ser/Thr protein kinase family. TGFB receptor subfamily.,similarity:Contains 1 GS domain.,similarity:Contains 1 protein kinase domain.,