



Rabbit Anti-BMPR1B antibody

SL6639R

Product Name:	BMPR1B
Chinese Name:	骨形态发生蛋白受体1B抗体
Alias:	BMPR-IB; Activin receptor like kinase 6; Acvrlk6; ALK 6; ALK6; alk6tr; BMP type-1B receptor; BMPR IB; BMPR-1B; Bmpr1b; BMPRIB; BMR1B_HUMAN; Bone morphogenetic protein receptor type 1B; Bone morphogenetic protein receptor type IB; Bone morphogenetic protein receptor type-1B; BR 1b; BR1b; CDw 293; CDw293; CDw293 antigen; CFK 43a; CFK43a; Serine/threonine receptor kinase; zALK 6; zALK6.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Cow,Rabbit,Sheep,.
Applications:	WB=1:500-2000ELISA=1:500-1000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	56kDa
Cellular localization:	The cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human BMPR1B:61-160/502<Extracellular>
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	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 BMP7/OP-1 and GDF5.

Involvement in disease; Defects in *BMPR1B* are the cause of acromesomelic chondrodysplasia with genital anomalies (AMDGA). 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).

Defects in *BMPR1B* are a cause of brachydactyly type A2 (BDA2) [MIM:112600].

Brachydactyly (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.

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 BMP7/OP-1 and GDF5.

Subcellular Location:

Membrane; Single-pass type I membrane protein.

DISEASE:

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Similarity:

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

Contains 1 GS domain.

Contains 1 protein kinase domain.

SWISS:
O00238

Gene ID:
658

Database links:

[Entrez Gene: 658](#)Human

[Entrez Gene: 12167](#)Mouse

[Omim: 603248](#)Human

[SwissProt: O00238](#)Human

[SwissProt: P36898](#)Mouse

[Unigene: 598475](#)Human

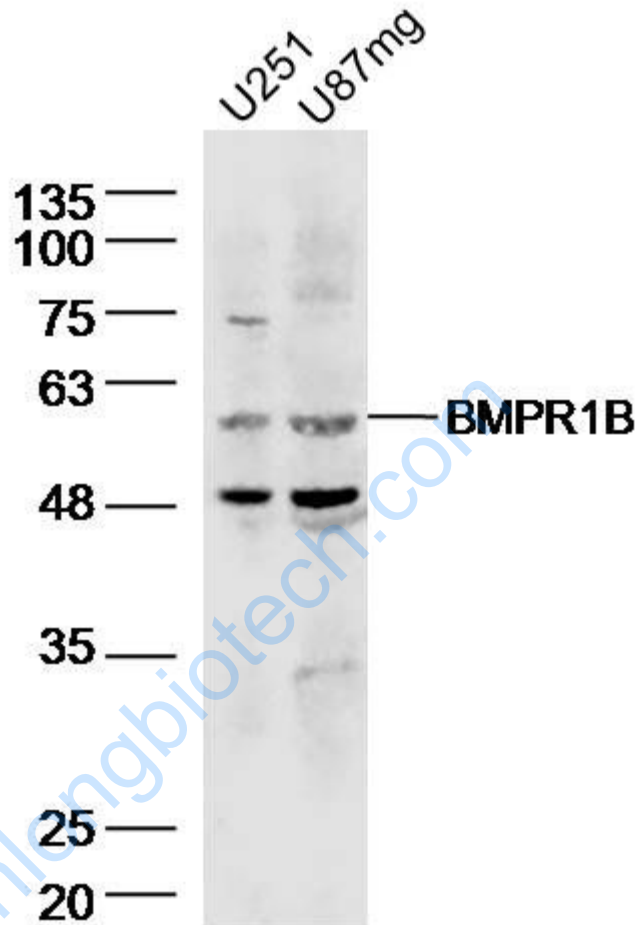
[Unigene: 39089](#)Mouse

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

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Picture:



Sample:

U251(human)cell Lysate at 30 ug

U87mg(human)cell Lysate at 30 ug

Primary: Anti- BMPR1B (SL6639R)at 1/500 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 54kD

Observed band size: 56 kD