### bs-6639R

## [ Primary Antibody ]



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# **BMPR1B** Rabbit pAb

DATASHEET -

Host: Rabbit Isotype: IgG

Clonality: Polyclonal

GeneID: 658 **SWISS:** 000238

Target: BMPR1B

**Immunogen:** KLH conjugated synthetic peptide derived from human BMPR1B:

61-160/502. < Extracellular >

Purification: affinity purified by Protein A

Concentration: 1mg/ml

Storage: 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50%

Glycerol.

Shipped at 4°C. Store at -20°C for one year. Avoid repeated

freeze/thaw cycles.

**Background:** 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]. 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.

Applications: WB (1:500-2000)

Reactivity: Human (predicted: Mouse,

Rat, Rabbit, Sheep, Cow,

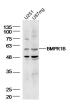
Dog)

Predicted 56 kDa

MW.:

Subcellular Cell membrane

### VALIDATION IMAGES



Sample: U251(human)cell Lysate at 30 ug U87mg(human)cell Lysate at 30 ug Primary: Anti-BMPR1B (bs-6639R)at 1/500 dilution Secondary: IRDve800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 54kD Observed band size: 56 kD

#### — SELECTED CITATIONS —

• [IF=3.688] Jianshu Lv. et al. Regulatory role of dihydrotestosterone on BMP-6 receptors in granular cells of sheep antral