

RPCB1172

Product Information

Product SKU:	RPCB1172	Gene ID:	659	Size:	10µg
Tag:	C-hFc	Reactivity:	Human		

Additional Information

Expression Host:	HEK293 cells	Swissprot:	Q13873-1
Purity:	> 95% by SDS-PAGE.		

Protein Information

Background: The bone morphogenetic protein type II receptor (BMPR-II, or BMPR2), a receptor for the transforming growth factor (TGF)-beta/bone morphogenetic protein (BMP) superfamily. Reduced expression or function of BMPR2 signaling leads to exaggerated TGF-beta signaling and altered cellular responses to TGF-beta. In endothelial cells, BMPR2 mutation increases the susceptibility of cells to apoptosis. BMPR2 transduces BMP signals by forming heteromeric complexes with and phosphorylating BMP type I receptors. The intracellular domain of BMPR2 is both necessary and sufficient for receptor complex interaction. It had been identified that BMPR2 plays a key role in cell growth. Its mutations lead to hereditary pulmonary hypertension, and knockout of Bmpr-II results in early embryonic lethality. The C-terminal tail of BMPR2 provides binding sites for a number of regulatory proteins that may initiate Smad-independent signalling. BMPR2 mutations were predicted to alter the BMP and TGF-b1/SMAD signalling pathways, resulting in proliferation rather than apoptosis of vascular cells, and greatly increase the risk of developing severe pulmonary arterial hypertension. BMPR2 gene result in familial Primary pulmonary hypertension (PPH) transmitted as an autosomal dominant trait, albeit with low penetrance. Heterozygous germline mutations of BMPR2 gene have been identified in patients with familial and sporadic PPH, indicating that BMPR2 may contribute to the maintenance of normal pulmonary vascular structure and function. Tctex-1, a light

chain of the motor complex dynein, interacts with the cytoplasmic domain of BMPR2 and demonstrate that Tctex-1 is phosphorylated by BMPR-II, a function disrupted by PPH disease causing mutations within exon 12. BMPR2 and Tctex-1 co-localize to endothelium and smooth muscle within the media of pulmonary arterioles, key sites of vascular remodelling in PPH.

- Protein Description:** High quality, high purity and low endotoxin recombinant Recombinant Human BMPR-2 Protein , tested reactivity in HEK293 cells and has been validated in SDS-PAGE.100% guaranteed.
- Endotoxin:** < 1 EU/μg of the protein by LAL method.
- Formulation:** Lyophilized from a 0.22 μm filtered solution of PBS, pH 7.4.
- Storage:** Store at -20°C.Store the lyophilized protein at -20°C to -80 °C up to 1 year from the date of receipt.After reconstitution, the protein solution is stable at -20°C for 3 months, at 2-8°C for up to 1 week.