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Recombinant Human BMPR2 / BMPR-II Protein (His & Fc tag)

产品货号: TP08454

第三版

别名: BMPR-II; BMPR3; BMR2; BRK-3; POVD1; PPH1; T-ALK

描述: 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.

配方: PBS

分子量: 42 kDa

序列: Met 1-Ile 151

纯度: > 95% by HPLC

浓度:

内毒素: <1.0 EU per 1 ug of protein (determined by LAL method)

存储: +4 ° C 保存 (1-2 周). 长期保存在 -20 ° C 或者 -70 ° C. 避免反复冻融.