

### Recombinant human BMPR2 protein ab126926

#### 5 图像

#### 描述

产品名称	重组人BMPR2蛋白
生物活性	The specific activity of ab126926 was determined to be 14 nmol/min/mg.
纯度	> 75 % Densitometry. Affinity purified.
表达系统	Baculovirus infected Sf9 cells
Accession	<b><u>Q13873</u></b>
蛋白长度	Protein fragment
无动物成分	No
性质	Recombinant
种属	Human
预测分子量	115 kDa including tags
氨基酸	174 to 1038
标签	His tag N-Terminus

#### 技术指标

Our **Abpromise guarantee** covers the use of **ab126926** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

应用	SDS-PAGE Functional Studies Western blot
形式	Liquid
补充说明	<b><u>ab64311</u></b> (Myelin Basic Protein protein) can be utilized as a substrate for assessing kinase activity

#### 制备和贮存

稳定性和存储	Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles. pH: 7.00 Preservative: 1.02% Imidazole
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Constituents: 0.002% PMSF, 0.81% Sodium phosphate, 0.0038% DTT, 25% Glycerol (glycerin, glycerine), 1.76% Sodium chloride

This product is an active protein and may elicit a biological response in vivo, handle with caution.

## 常规信息

### 功能

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. Binds to BMP-7, BMP-2 and, less efficiently, BMP-4. Binding is weak but enhanced by the presence of type I receptors for BMPs.

### 组织特异性

Highly expressed in heart and liver.

### 疾病相关

Defects in BMPR2 are the cause of primary pulmonary hypertension (PPH1) [MIM:178600]. PPH1 is a rare autosomal dominant disorder characterized by plexiform lesions of proliferating endothelial cells in pulmonary arterioles. The lesions lead to elevated pulmonary arterial pressure, right ventricular failure, and death. The disease can occur from infancy throughout life and it has a mean age at onset of 36 years. Penetrance is reduced. Although familial PPH1 is rare, cases secondary to known etiologies are more common and include those associated with the appetite-suppressant drugs.

Defects in BMPR2 are a cause of pulmonary venoocclusive disease (PVOD) [MIM:265450]. PVOD is a rare form of pulmonary hypertension in which the vascular changes originate in the small pulmonary veins and venules. The pathogenesis is unknown and any link with PPH1 has been speculative. The finding of PVOD associated with a BMPR2 mutation reveals a possible pathogenetic connection with PPH1.

### 序列相似性

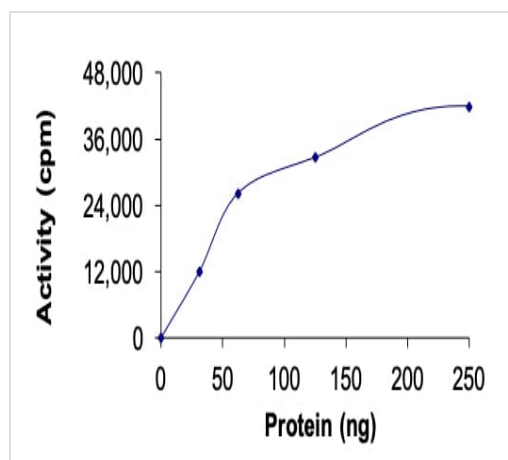
Belongs to the protein kinase superfamily. TKL Ser/Thr protein kinase family. TGFB receptor subfamily.

Contains 1 protein kinase domain.

### 细胞定位

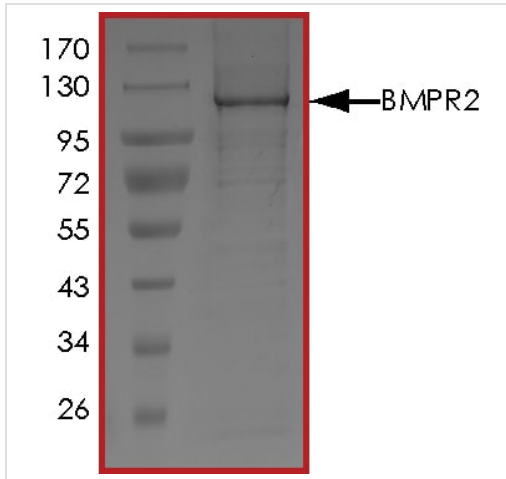
Membrane.

## 图片



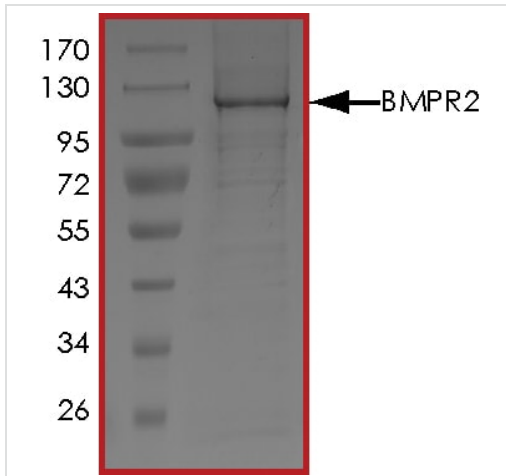
The specific activity of BMPR2 (ab126926) was determined to be 11.9 nmol/min/mg as per activity assay protocol

Functional Studies - Recombinant human BMPR2 protein (ab126926)



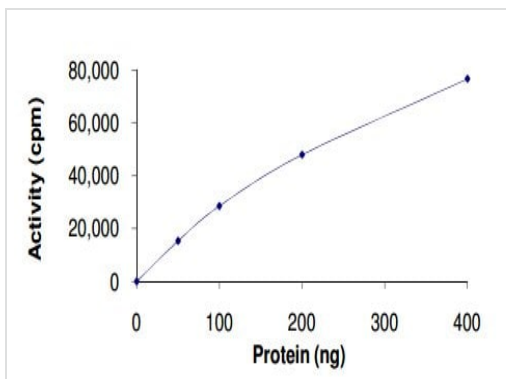
SDS PAGE analysis of ab126926

SDS-PAGE - Recombinant human BMPR2 protein  
(ab126926)



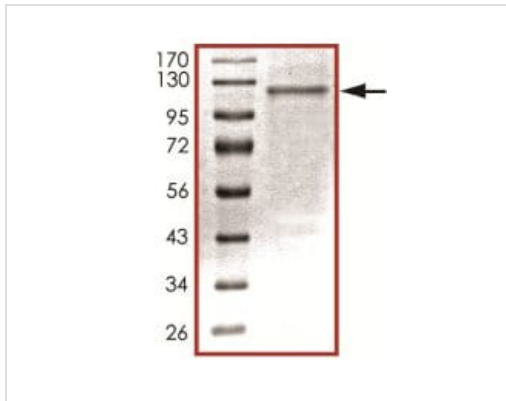
SDS PAGE analysis of ab126926

SDS-PAGE - Recombinant human BMPR2 protein  
(ab126926)



The specific activity of ab126926 was determined to be 14 nmol/min/mg.

Functional Studies - Recombinant human BMPR2  
protein (ab126926)



SDS-PAGE analysis of ab126926.

SDS-PAGE - Recombinant human BMPR2 protein  
(ab126926)

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