# abcam

# Product datasheet

# Recombinant human BMPR2 protein ab126926

# 5 Images

**Description** 

Product name Recombinant human BMPR2 protein

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

**Purity** > 75 % Densitometry.

Affinity purified.

Expression system Baculovirus infected Sf9 cells

Accession Q13873

Protein length Protein fragment

Animal free No

Nature Recombinant

**Species** Human

Predicted molecular weight 115 kDa including tags

Amino acids 174 to 1038

Tags His tag N-Terminus

# **Specifications**

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.

Applications SDS-PAGE

**Functional Studies** 

Western blot

Form Liquid

Additional notes <u>ab64311</u> (Myelin Basic Protein protein) can be utilized as a substrate for assessing kinase

activity

**Preparation and Storage** 

Stability and Storage 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.

#### **General Info**

#### **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. 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.

#### Tissue specificity

#### Involvement in disease

Highly expressed in heart and liver.

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 pression, 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.

### Sequence similarities

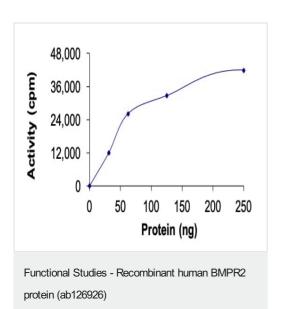
Belongs to the protein kinase superfamily. TKL Ser/Thr protein kinase family. TGFB receptor subfamily.

Contains 1 protein kinase domain.

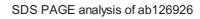
#### **Cellular localization**

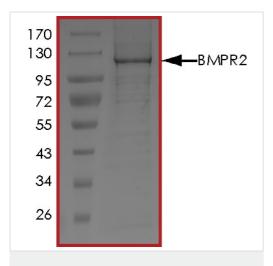
Membrane.

#### **Images**

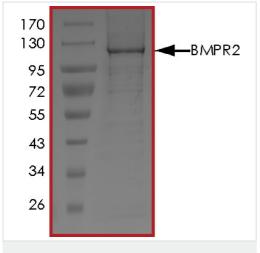


The specific activity of BMPR2 (ab126926) was determined to be 11.9 nmol/min/mg as per activity assay protocol

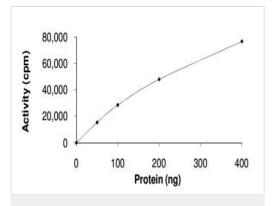




SDS-PAGE - Recombinant human BMPR2 protein (ab126926)



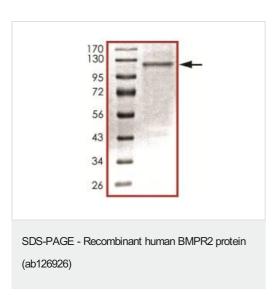
SDS-PAGE - Recombinant human BMPR2 protein (ab126926)



Functional Studies - Recombinant human BMPR2 protein (ab126926)

SDS PAGE analysis of ab126926

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



SDS-PAGE analysis of ab126926.

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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