

## Rabbit Anti-ACVRL1/AP Conjugated antibody

SL6020R-AP

<b>Product Name</b>	Anti-ACVRL1/AP
<b>Chinese Name</b>	碱性磷酸酶（AP）标记的激活素受体样激酶 1 抗体
<b>Alias</b>	Activin A receptor; Activin A receptor type II like 1; Activin receptor like kinase 1; Activin receptor-like kinase 1; ACVL1_HUMAN; ACVRL1; ACVRLK1; ALK-1; ALK1; HHT; HHT2; ORW2; Osler Rendu Weber syndrome 2; Serine/threonine protein kinase receptor R3; Serine/threonine-protein kinase receptor R3; SKR3; TGF B superfamily receptor type I; TGF-B superfamily receptor type I; TSR-I; TSR1.
<b>Research Area</b>	Cardiovascular Cell biology immunology Signal transduction transcriptional regulatory factor Kinases and Phosphatases Cell Surface Molecule
<b>Immunogen Species</b>	Rabbit
<b>Clonality</b>	Polyclonal
<b>React Species</b>	Human,Mouse,Rat(predicted:Dog,Pig,Horse) IHC-P=1:100-500,IHC-F=1:100-500
<b>Applications</b>	not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
<b>Molecular weight</b>	53kDa
<b>Form</b>	Lyophilized or Liquid
<b>Concentration</b>	1mg/ml
<b>immunogen</b>	KLH conjugated synthetic peptide derived from human ACVRL1/ALK1
<b>Lsotype</b>	IgG
<b>Purification</b>	affinity purified by Protein A
<b>Storage Buffer</b>	1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
<b>Storage</b>	
<b>Product Detail</b>	<b>background:</b> 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 TGF-beta. May bind activin as well.

**Function:**

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**Subcellular Location:**

Membrane.

**DISEASE:**

Defects in ACVRL1 are the cause of hereditary hemorrhagic telangiectasia type 2 (HHT2) [MIM:600376]; also known as Osler-Rendu-Weber syndrome 2 (ORW2). HHT2 is an autosomal dominant multisystemic vascular dysplasia, characterized by recurrent epistaxis, muco-cutaneous telangiectases, gastro-intestinal hemorrhage, and pulmonary, cerebral and hepatic arteriovenous malformations; all secondary manifestations of the underlying vascular dysplasia.

**Similarity:**

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

Contains 1 GS domain.

Contains 1 protein kinase domain.

**Database links:**

[Entrez Gene: 94](#) Human

[Omim: 601284](#) Human

[SwissProt: P37023](#) Human

[Unigene: 591026](#) Human

**Important Note:**

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.