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ACVRL1 Rabbit pAb

Catalog # AP99342

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P37023
Other Accession	P37023
Reactivity	Rat, Human, Mouse
Predicted	Pig, Dog, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	56124
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human ACVRL1
Epitope Specificity	21-120/503
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
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.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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.

Additional Information

Gene ID	94
Other Names	ACVRLK1; ALK-1; ALK1; HHT; HHT2; ORW2; SKR3; TSR-I; ACVL1_HUMAN; ACVRL1; Activin receptor-like kinase 1 (ALK-1); Serine/threonine-protein kinase receptor R3 (SKR3); TGF-B superfamily receptor type I (TSR-I); 2.7.11.30; activin A receptor like type 1; activin A receptor type II-like 1; activin A receptor type IL; activin receptor-like kinase 1
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500

Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	ACVRL1
Synonyms	ACVRLK1, ALK1
Function	Type I receptor for TGF-beta family ligands BMP9/GDF2 and BMP10 and important regulator of normal blood vessel development. 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. May bind activin as well.
Cellular Location	Cell membrane; Single-pass type I membrane protein

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.