Revision 1

Store at -20C

#4398

ACVR1 Antibody

Cell Signaling TECHNOLOGY®

Orders: 877-616-CELL (2355)
orders@cellsignal.com

Support: 877-678-TECH (8324)

Web: info@cellsignal.com
cellsignal.com

3 Trask Lane | Danvers | Massachusetts | 01923 | USA

For Research Use Only. Not for Use in Diagnostic Procedures.

Applications:	Reactivity:	Sensitivity:	MW (kDa):	Source/Isotype:	UniProt ID:	Entrez-Gene Id:
W	H M Mk	Endogenous	57	Rabbit	#Q04771	90

Product Usage Information

Application	Dilution
Western Blotting	1:1000

Storage

Supplied in 10 mM sodium HEPES (pH 7.5), 150 mM NaCl, 100 µg/ml BSA and 50% glycerol. Store at –20°C. Do not aliquot the antibody.

Specificity/Sensitivity

ACVR1 Antibody detects endogenous levels of total ACVR1 protein. The antibody also cross-reacts with 35 kDa and 95 kDa bands of unknown origin.

Source / Purification

Polyclonal antibodies are produced by immunizing animals with a synthetic peptide corresponding to residues near the carboxy terminus of human ACVR1 protein. Antibodies were purified by protein A and peptide affinity chromatography.

Background

Activin receptor type 1 (ACVR1), also known as Activin receptor-like kinase 2 (ALK2) is a membrane associated serine/threonine kinase receptor that binds activin A and related TGF-β superfamily growth factors, including bone morphogenetic protein 2 (BMP2), BMP4, BMP6, and BMP7 (1-3). Functional BMP receptor tetramers are composed of a pair of type I (ACVR1, ACVRL1, BMPR1A, or BMPR1B) and a pair of type II (BMPR2, ACVR2A, ACVR2B) receptor proteins (4). Following ligand binding, ACVR1 is phosphorylated by its type II binding partner, which allows recruitment and activation of receptor regulated SMAD (R-Smad) proteins (5). Phosphorylation of R-Smad proteins results in their translocation to the nucleus where R-Smads stimulate transcription of target genes (6). Mutations in the corresponding ACVR1 gene, including R206H and Q207D, cause constitutive activation of the receptor, and are associated with the genetic developmental disorders hereditary hemorrhagic telangiectasia and fibrodysplasia ossificans progressive (FOP) (7-11). In a mouse model of FOP, overexpression of the wild-type ACVR1 gene lessened the skeletal lesions associated with the R206H genetic background (12). Small molecule inhibitors of the ACVR1 kinase domain can disrupt downstream SMAD1/5 activation, offering a promising therapeutic avenue in FOP, cancers, or other disorders arising from gain-of-function ACVR1 mutations (13,14).

Background References

1. Attisano, L. et al. (1993) *Cell* 75, 671-80.
2. Macías-Silva, M. et al. (1998) *J Biol Chem* 273, 25628-36.
3. Ho, C.C. and Bernard, D.J. (2009) *Biol Reprod* 81, 133-41.
4. Agnew, C. et al. (2021) *Nat Commun* 12, 4950.
5. Attisano, L. et al. (1996) *Mol Cell Biol* 16, 1066-73.
6. Attisano, L. and Wrana, J.L. (2000) *Curr Opin Cell Biol* 12, 235-43.
7. Shore, E.M. et al. (2006) *Nat Genet* 38, 525-7.
8. Letteboer, T.G. et al. (2005) *Hum Genet* 116, 8-16.
9. Song, G.A. et al. (2010) *J Biol Chem* 285, 22542-53.
10. Pan, H. et al. (2020) *J Musculoskelet Neuronal Interact* 20, 149-159.
11. Ramachandran, A. et al. (2021) *EMBO J* 40, e106317.
12. Yamamoto, M. et al. (2022) *J Bone Miner Res* 37, 2077-2093.
13. Sanvitale, C.E. et al. (2013) *PLoS One* 8, e62721.
14. Jimmidi, R. et al. (2024) *Proc Natl Acad Sci U S A* 121, e2413108121.

Species Reactivity

Species reactivity is determined by testing in at least one approved application (e.g., western blot).

Western Blot Buffer

IMPORTANT: For western blots, incubate membrane with diluted primary antibody in 5% w/v BSA, 1X TBS, 0.1% Tween® 20 at 4°C with gentle shaking, overnight.

Applications Key

W: Western Blotting

Cross-Reactivity Key

H: Human **M:** Mouse **Mk:** Monkey

Trademarks and Patents

Cell Signaling Technology is a trademark of Cell Signaling Technology, Inc.

All other trademarks are the property of their respective owners. Visit cellsignal.com/trademarks for more information.

Limited Uses

Except as otherwise expressly agreed in a writing signed by a legally authorized representative of CST, the following terms apply to Products provided by CST, its affiliates or its distributors. Any Customer's terms and conditions that are in addition to, or different from, those contained herein, unless separately accepted in writing by a legally authorized representative of CST, are rejected and are of no force or effect.

Products are labeled with For Research Use Only or a similar labeling statement and have not been approved, cleared, or licensed by the FDA or other regulatory foreign or domestic entity, for any purpose. Customer shall not use any Product for any diagnostic or therapeutic purpose, or otherwise in any manner that conflicts with its labeling statement. Products sold or licensed by CST are provided for Customer as the end-user and solely for research and development uses. Any use of Product for diagnostic, prophylactic or therapeutic purposes, or any purchase of Product for resale (alone or as a component) or other commercial purpose, requires a separate license from CST. Customer shall (a) not sell, license, loan, donate or otherwise transfer or make available any Product to any third party, whether alone or in combination with other materials, or use the Products to manufacture any commercial products, (b) not copy, modify, reverse engineer, decompile, disassemble or otherwise attempt to discover the underlying structure or technology of the Products, or use the Products for the purpose of developing any products or services that would compete with CST products or services, (c) not alter or remove from the Products any trademarks, trade names, logos, patent or copyright notices or markings, (d) use the Products solely in accordance with CST Product Terms of Sale and any applicable documentation, and (e) comply with any license, terms of service or similar agreement with respect to any third party products or services used by Customer in connection with the Products.