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EVC2 (A-3): sc-515747

BACKGROUND

Ellis van Creveld syndrome 2 (EVC2), also designated limbin, is a protein containing a leucine zipper and a transmembrane domain. EVC2 is expressed in the developing vertebral bodies, kidney, ribs, lung, upper and lower limbs and heart. This protein is implicated in two major diseases: Ellis-van Creveld syndrome (EVC) and Weyers acrodermal dysostosis (WAD). EVC is characterized by short-limb dwarfism, short ribs and dysplastic nails and teeth. It is an autosomal recessive disorder often causing heart defects. WAD is an autosomal dominant disorder and although the phenotype of WAD is milder than EVC, it still causes dysplastic nails, short limbs and short stature.

REFERENCES

1. Ide, S.E., et al. 1996. Exclusion of the MSX1 homeobox gene as the gene for the Ellis van Creveld syndrome in the Amish. *Hum. Genet.* 98: 572-575.
2. Brueton, L.A., et al. 1990. Ellis-van creveld syndrome, Jeune syndrome, and renal-hepatic-pancreatic dysplasia: separate entities or disease spectrum? *J. Med. Genet.* 27: 252-255.
3. McKusick, V.A. 2000. Ellis-van Creveld syndrome and the Amish. *Nat. Genet.* 24: 203-204.
4. Tompson, S.W., et al. 2001. Ellis-van Creveld syndrome resulting from segmental uniparental disomy of chromosome 4. *J. Med. Genet.* 38: E18.
5. Arya, L., et al. 2001. Ellis-van Creveld Syndrome: a report of two cases. *Pediatr. Dermatol.* 18: 485-489.
6. Galdzicka, M., et al. 2002. A new gene, EVC2, is mutated in Ellis-van Creveld syndrome. *Mol. Genet. Metab.* 77: 291-295.
7. Sajeev, C.G., et al. 2002. Images in cardiology: common atrium in a child with Ellis-Van Creveld syndrome. *Heart* 88: 142.

CHROMOSOMAL LOCATION

Genetic locus: EVC2 (human) mapping to 4p16.2; Evc2 (mouse) mapping to 5 B3.

SOURCE

EVC2 (A-3) is a mouse monoclonal antibody raised against amino acids 370-508 mapping within an internal region of EVC2 of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

EVC2 (A-3) is available conjugated to agarose (sc-515747 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-515747 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-515747 PE), fluorescein (sc-515747 FITC), Alexa Fluor[®] 488 (sc-515747 AF488), Alexa Fluor[®] 546 (sc-515747 AF546), Alexa Fluor[®] 594 (sc-515747 AF594) or Alexa Fluor[®] 647 (sc-515747 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor[®] 680 (sc-515747 AF680) or Alexa Fluor[®] 790 (sc-515747 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

EVC2 (A-3) is recommended for detection of EVC2 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for EVC2 siRNA (h): sc-105339, EVC2 siRNA (m): sc-144961, EVC2 shRNA Plasmid (h): sc-105339-SH, EVC2 shRNA Plasmid (m): sc-144961-SH, EVC2 shRNA (h) Lentiviral Particles: sc-105339-V and EVC2 shRNA (m) Lentiviral Particles: sc-144961-V.

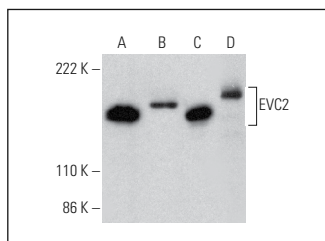
Molecular Weight of EVC2: 153 kDa.

Positive Controls: U-698-M whole cell lysate: sc-364799, A549 cell lysate: sc-2413 or NIH/3T3 whole cell lysate: sc-2210.

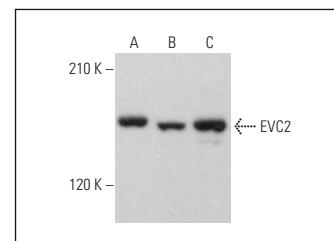
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker[™] Molecular Weight Standards: sc-2035, UltraCruz[®] Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz[®] Mounting Medium: sc-24941 or UltraCruz[®] Hard-set Mounting Medium: sc-359850.

DATA



EVC2 (A-3): sc-515747. Western blot analysis of EVC2 expression in U-698-M (A), F9 (B) and SK-MEL-28 (C) whole cell lysates and human testis tissue extract (D).



EVC2 (A-3): sc-515747. Western blot analysis of EVC2 expression in A549 (A), Neuro-2A (B) and NIH/3T3 (C) whole cell lysates.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.