

EVC (C-6): sc-377157

BACKGROUND

EVC, or Ellis-van Creveld syndrome, is an autosomal skeletal dysplasia caused by mutations in the EVC and EVC2 genes. Found in developing ribs, heart, kidney and lung, the EVC gene is responsible for normal development of the face, limbs, teeth and nails. The protein expressed by the EVC gene is an intracellular component of the hedgehog signal pathway that contains a leucine zipper and transmembrane domain. Defects in the EVC gene can lead to short-limb dwarfism, ectodermal dysplasia and cardiac anomalies such as irregular atrioventricular septum development. Additionally, the EVC gene has been implicated in Weyers acrodermal dysostosis, an autosomal dominant disease characterized by facial abnormalities and limb defects.

REFERENCES

1. Polymeropoulos, M.H., et al. 1996. The gene for the Ellis-van Creveld syndrome is located on chromosome 4p16. *Genomics* 35: 1-5.
2. Ruiz-Perez, V.L., et al. 2000. Mutations in a new gene in Ellis-van Creveld syndrome and Weyers acrodermal dysostosis. *Nat. Genet.* 24: 283-286.
3. Galdzicka, M., et al. 2002. A new gene, EVC is mutated in Ellis-van Creveld syndrome. *Mol. Genet. Metab.* 77: 291-295.
4. Mostafa, M.I., et al. 2005. Unusual pattern of inheritance and orodental changes in the Ellis-van Creveld syndrome. *Genet. Couns.* 16: 75-83.
5. Scurlock, D., et al. 2005. Ellis-van Creveld syndrome and dyserythropoiesis. *Arch. Pathol. Lab. Med.* 129: 680-682.
6. van Hagen, J.M., et al. 2005. From gene to disease; EVC, EVC2, and Ellis-van Creveld syndrome. *Ned. Tijdschr. Geneesk.* 149: 929-931.
7. Ye, X., et al. 2006. A novel heterozygous deletion in the EVC2 gene causes Weyers acrofacial dysostosis. *Hum. Genet.* 119: 199-205.
8. Ruiz-Perez, V.L., et al. 2007. EVC is a positive mediator of Ihh-regulated bone growth that localises at the base of chondrocyte cilia. *Development* 134: 2903-2912.

CHROMOSOMAL LOCATION

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

SOURCE

EVC (C-6) is a mouse monoclonal antibody raised against amino acids 181-441 mapping within an internal region of EVC of mouse 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.

STORAGE

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

APPLICATIONS

EVC (C-6) is recommended for detection of EVC 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), immunohistochemistry (including paraffin-embedded sections) (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 EVC siRNA (h): sc-72235, EVC siRNA (m): sc-72236, EVC shRNA Plasmid (h): sc-72235-SH, EVC shRNA Plasmid (m): sc-72236-SH, EVC shRNA (h) Lentiviral Particles: sc-72235-V and EVC shRNA (m) Lentiviral Particles: sc-72236-V.

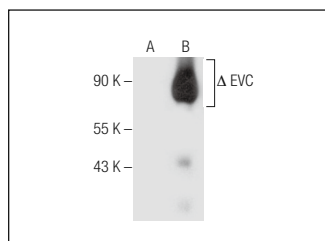
Molecular Weight of EVC: 124 kDa.

Positive Controls: EVC (m): 293T Lysate: sc-120134.

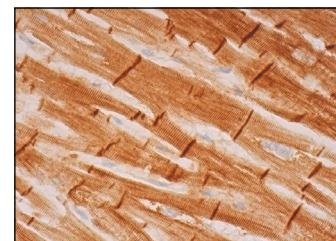
RECOMMENDED SUPPORT REAGENTS

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. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



EVC (C-6): sc-377157. Western blot analysis of EVC expression in non-transfected: sc-117752 (A) and truncated mouse EVC transfected: sc-120134 (B) 293T whole cell lysates.



EVC (C-6): sc-377157. Immunoperoxidase staining of formalin fixed, paraffin-embedded human heart muscle tissue showing membrane and cytoplasmic staining of myocytes.

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.