

SHFM3 (H-300): sc-292027

BACKGROUND

SHFM3 (split-hand/foot malformation type 3), also known as FBXW4 (F-box and WD-40 domain-containing protein 4) or Dactylin, is a 412 amino acid protein that is primarily involved in signaling pathways that are essential for normal limb development. Specifically, SHFM3 promotes ubiquitination and degradation of certain phosphorylated proteins. Defects in the gene encoding SHFM3 are the cause of split-hand/foot malformation type 3, an autosomal dominant disorder that is characterized by hypoplasia/aplasia of the central digits, causing variable fusion with the remaining digits. The disease is not a result of a point mutation, but rather a genomic rearrangement resulting in a tandem duplication that contains a disrupted copy of the gene encoding SHFM3, as well as extra copies of two other genes that are linked to limb development. SHFM3 is typically expressed in kidney, brain, lung and liver, with highest expression in fetal brain.

REFERENCES

1. Ianakiev, P., et al. 1999. A novel human gene encoding an F-box/WD40 containing protein maps in the SHFM3 critical region on 10q24. *Biochem. Biophys. Res. Commun.* 261: 64-70.
2. Sidow, A., et al. 1999. A novel member of the F-box/WD40 gene family, encoding dactylin, is disrupted in the mouse dactylaplasia mutant. *Nat. Genet.* 23: 104-107.
3. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 608071. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
4. Basel, D., et al. 2003. Split hand foot malformation is associated with a reduced level of dactylin gene expression. *Clin. Genet.* 64: 350-354.
5. de Mollerat, X.J., et al. 2003. A genomic rearrangement resulting in a tandem duplication is associated with split hand-split foot malformation 3 (SHFM3) at 10q24. *Hum. Mol. Genet.* 12: 1959-1971.
6. Kano, H., et al. 2005. Genomic rearrangement at 10q24 in non-syndromic split-hand/split-foot malformation. *Hum. Genet.* 118: 477-483.

CHROMOSOMAL LOCATION

Genetic locus: FBXW4 (human) mapping to 10q24.32; Fbxw4 (mouse) mapping to 19 C3.

SOURCE

SHFM3 (H-300) is a rabbit polyclonal antibody raised against amino acids 113-412 mapping at the C-terminus of SHFM3 of human origin.

PRODUCT

Each vial contains 200 µg IgG 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

SHFM3 (H-300) is recommended for detection of SHFM3 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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).

SHFM3 (H-300) is also recommended for detection of SHFM3 in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for SHFM3 siRNA (h): sc-90449, SHFM3 siRNA (m): sc-153450, SHFM3 shRNA Plasmid (h): sc-90449-SH, SHFM3 shRNA Plasmid (m): sc-153450-SH, SHFM3 shRNA (h) Lentiviral Particles: sc-90449-V and SHFM3 shRNA (m) Lentiviral Particles: sc-153450-V.

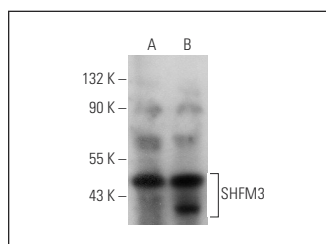
Molecular Weight of SHFM3: 46 kDa.

Positive Controls: mouse lung extract: sc-2390 or mouse heart extract: sc-2254.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



SHFM3 (H-300): sc-292027. Western blot analysis of SHFM3 expression in mouse lung (A) and mouse heart (B) tissue extracts.

RESEARCH USE

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

PROTOCOLS

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