

# MCFD2 (Z-7): sc-101281

## BACKGROUND

Multiple coagulation factor deficiency protein 2 (MCFD2) is localized in the endoplasmic reticulum-Golgi intermediate compartment (ERGIC) through a direct, calcium-dependent interaction with LMAN1. The MCFD2-LMAN1 complex forms a specific cargo receptor for the transport of selected proteins from the endoplasmic reticulum to the Golgi apparatus. Mutations in the MCFD2 gene may cause Factor V and Factor VIII combined deficiency (F5F8D). F5F8D is an autosomal recessive human bleeding disorder characterized by the reduction of both clotting proteins.

## REFERENCES

- Zhang, B., et al. 2003. Bleeding due to disruption of a cargo-specific ER-to-Golgi transport complex. *Nat. Genet.* 34: 220-225.
- Spatuzza, C., et al. 2004. Heat shock induces preferential translation of ERGIC-53 and affects its recycling pathway. *J. Biol. Chem.* 279: 42535-42544.
- Zhang, B., et al. 2004. Familial multiple coagulation factor deficiencies: new biologic insight from rare genetic bleeding disorders. *J. Thromb. Haemost.* 2: 1564-1572.
- Zhang, B., et al. 2005. LMAN1 and MCFD2 form a cargo receptor complex and interact with coagulation Factor VIII in the early secretory pathway. *J. Biol. Chem.* 280: 25881-25886.
- Mohanty, D., et al. 2005. Mutations in the MCFD2 gene and a novel mutation in the LMAN1 gene in Indian families with combined deficiency of Factor V and VIII. *Am. J. Hematol.* 79: 262-266.
- Zhang, B., et al. 2005. Combined deficiency of factor V and factor VIII is due to mutations in either LMAN1 or MCFD2. *Blood* 107: 1903-1907.

## CHROMOSOMAL LOCATION

Genetic locus: MCFD2 (human) mapping to 2p21.

## SOURCE

MCFD2 (Z-7) is a mouse monoclonal antibody raised against recombinant MCFD2 of human origin.

## PRODUCT

Each vial contains 100 µg IgG<sub>2a</sub> 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.

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) for detailed protocols and support products.

## APPLICATIONS

MCFD2 (Z-7) is recommended for detection of MCFD2 of human origin by 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 MCFD2 siRNA (h): sc-44445, MCFD2 shRNA Plasmid (h): sc-44445-SH and MCFD2 shRNA (h) Lentiviral Particles: sc-44445-V.

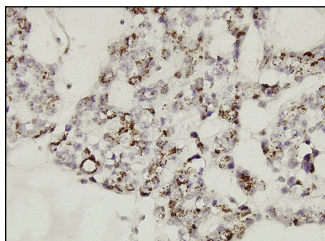
Molecular Weight of MCFD2: 17 kDa.

## RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended:

- Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz<sup>®</sup> Mounting Medium: sc-24941 or UltraCruz<sup>®</sup> Hard-set Mounting Medium: sc-359850.

## DATA



MCFD2 (Z-7): sc-101281. Immunoperoxidase staining of formalin-fixed, paraffin-embedded human ovary, clear cell carcinoma tissue showing cytoplasmic localization.

## RESEARCH USE

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