

MCFD2 (N-16): sc-46391

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 of 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

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- 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. 2006. 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 (N-16) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the N-terminus of MCFD2 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.

Blocking peptide available for competition studies, sc-46391 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

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 or our catalog for detailed protocols and support products.

APPLICATIONS

MCFD2 (N-16) is recommended for detection of MCFD2 of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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).

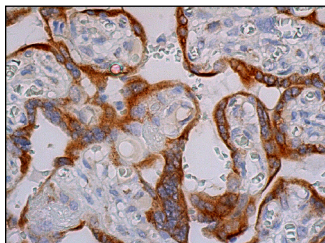
Suitable for use as control antibody for MCFD2 siRNA (h): sc-44445, MCFD2 shRNA Plasmid (h): sc-44445-SH and MCFD2 shRNA (h) Lentiviral Particles: sc-44445-V.

Molecular Weight of MCFD2: 17 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (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) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941. 3) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

DATA



MCFD2 (N-16): sc-46391. Immunoperoxidase staining of formalin fixed, paraffin-embedded human placenta tissue showing cytoplasmic staining of trophoblastic cells.

RESEARCH USE

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