

# FANCC (6E7): sc-293308

## BACKGROUND

Fanconi anemia (FA) is an autosomal recessive disorder characterized by bone marrow failure, birth defects and chromosomal instability. The FA Group C complementation group gene encodes the protein FANCC, which is located in both cytoplasmic and nuclear compartments. FANCC is expressed in a cell cycle-dependent manner, with the lowest levels at the G<sub>1</sub>/S boundary and the highest levels in the M-phase. The FANCC protein interacts with other FA complementation group proteins as well as non-FA proteins. A human a spectrin II (designated aSp11s) acts as a scaffold to enhance interactions between FANCC and FANCA to form a nuclear complex. Another binding partner of FANCC is the BTB/POZ domain containing protein FAZF, which is a transcriptional repressor. In hematopoietic cells expressing mutant FANCC, PKR is constitutively phosphorylated and has increased binding affinity for double-stranded RNA, which suggests that FANCC indirectly suppresses the activity of PKR. These cells are also apoptotic and are hypersensitive to IFN $\gamma$  and TNF $\alpha$ . In addition, FANCC protein is involved in the activation of STAT1 through receptors for at least three hematopoietic growth and survival factors.

## REFERENCES

- McMahon, L.W., et al. 1999. Human  $\alpha$  spectrin II and the Fanconi anemia proteins FANCA and FANCC interact to form a nuclear complex. *J. Biol. Chem.* 274: 32904-32908.
- Hoatlin, M.E., et al. 1999. A novel BTB/POZ transcriptional repressor protein interacts with the Fanconi anemia group C protein and PLZF. *Blood* 94: 3737-3747.
- Kruyt, F.A., et al. 1999. Resistance to mitomycin C requires interaction between the Fanconi anemia proteins FANCA and FANCC in the nucleus through an arginine-rich domain. *J. Biol. Chem.* 274: 34212-34218.
- Kupfer, G., et al. 1999. A patient-derived mutant form of the Fanconi anemia protein, FANCA, is defective in nuclear accumulation. *Exp. Hematol.* 27: 587-593.
- Heinrich, M.C., et al. 2000. Posttranscriptional cell cycle-dependent regulation of human FANCC expression. *Blood* 95: 3970-3977.
- Pang, Q., et al. 2000. The Fanconi anemia protein FANCC binds to and facilitates the activation of STAT1 by  $\gamma$  interferon and hematopoietic growth factors. *Mol. Cell. Biol.* 20: 4724-4735.
- Pang, Q., et al. 2001. Role of double-stranded RNA-dependent protein kinase in mediating hypersensitivity of Fanconi anemia complementation group C cells to interferon  $\gamma$ , tumor necrosis factor- $\alpha$ , and double stranded RNA. *Blood* 97: 1644-1652.

## CHROMOSOMAL LOCATION

Genetic locus: FANCC (human) mapping to 9q22.32.

## SOURCE

FANCC (6E7) is a mouse monoclonal antibody raised against amino acids 1-100 of FANCC of human origin.

## PRODUCT

Each vial contains 100  $\mu$ g IgG<sub>2b</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

## APPLICATIONS

FANCC (6E7) is recommended for detection of FANCC of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2  $\mu$ g per 100-500  $\mu$ g 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 FANCC siRNA (h): sc-35354, FANCC shRNA Plasmid (h): sc-35354-SH and FANCC shRNA (h) Lentiviral Particles: sc-35354-V.

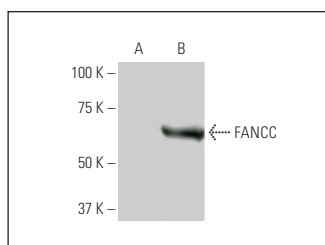
Molecular Weight of FANCC: 60 kDa.

Positive Controls: FANCC transfected 293T whole cell lysate.

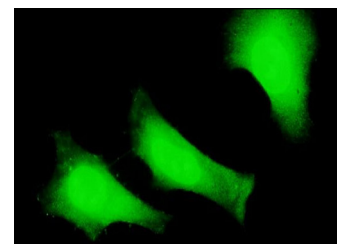
## RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker<sup>TM</sup> Molecular Weight Standards: sc-2035, UltraCruz<sup>®</sup> 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-IgG $\kappa$  BP-FITC: sc-516140 or m-IgG $\kappa$  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. 4) Immunohistochemistry: use m-IgG $\kappa$  BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

## DATA



FANCC (6E7): sc-293308. Western blot analysis of FANCC expression in non-transfected (A) and FANCC transfected (B) 293T whole cell lysates.



FANCC (6E7): sc-293308. Immunofluorescence staining of methanol-fixed HeLa cells showing cytoplasmic and nuclear localization.

## 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.