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Rabbit Anti-FANCC antibody

SL13140R

Product Name:	FANCC
Chinese Name:	范可尼综合征相关蛋白FANCC抗体
Alias:	bA80I15.1; FA 3; FA3; FAC; FACC; FANCC; FANCC_HUMAN; Fanconi anemia complementation group C; Fanconi anemia complementation group C protein; Fanconi anemia group C protein; Fanconi pancytopenia type 3; FLJ14675; Protein FACC.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Horse,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	63kDa
Cellular localization:	The nucleuscytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human FANCC:61-160/558
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	Fanconi anemia (FA) is an autosomal recessive disorder characterized by bone marrow failure, birth defects and chromsomal instability (1,2). 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 G1/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 (3). A human a spectrin II (designated aSpIIs) acts as a scaffold to enhance interactions between FANCC and FANCA to form a nuclear complex (4,5). Another binding partner of FANCC is the BTB/POZ domain containing protein FAZF, which is a transcriptional repressor (6). In hematopoietic cells expressing mutant FANCC, PKR is constitutively phosphorylated and has increased binding affinity for double-stranded RNA (7,8), which suggests that FANCC indirectly suppresses the activity of PKR. These cells are also apoptotic and are hypersensitive to IFNg and TNFa (8). In addition, FANCC protein is involved in the activation of STAT1 through receptors for at least three hematopoietic growth and survival factors (8).

Function:
DNA repair protein that may operate in a postreplication repair or a cell cycle checkpoint function. May be implicated in interstrand DNA cross-link repair and in the maintenance of normal chromosome stability. Upon IFNG induction, may facilitate STAT1 activation by recruiting STAT1 to IFNGR1.

Subunit:
Belongs to the multisubunit FA complex composed of FANCA, FANCB, FANCC, FANCE, FANCF, FANCG, FANCL/PHF9 and FANCM. This complex may also include HSP70. The complex is not found in FA patients. Interacts with ZBTB32. Upon IFNG induction, interacts with STAT1. Interacts with CDK1. Interacts with EIF2AK2; interaction between FA variants and EIF2AK2 may lead to augmented EIF2AK2 activation and cell death.

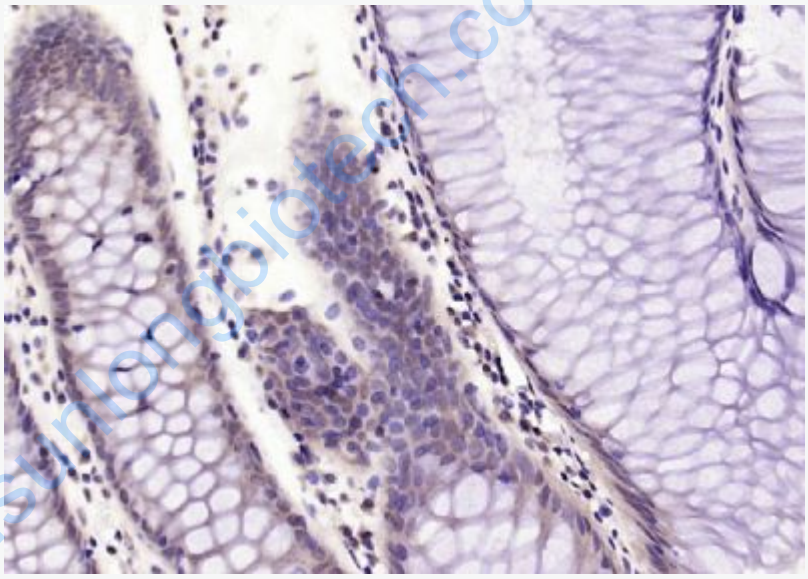
Subcellular Location:
Nucleus. Cytoplasm. The major form is nuclear. The minor form is cytoplasmic.

Tissue Specificity:
Ubiquitous.

DISEASE:
Defects in FANCC are the cause of Fanconi anemia complementation group C (FANCC) [MIM:227645]. A disorder affecting all bone marrow elements and resulting in anemia, leukopenia and thrombopenia. It is associated with cardiac, renal and limb malformations, dermal pigmentary changes, and a predisposition to the development of malignancies. At the cellular level it is associated with hypersensitivity to DNA-damaging agents, chromosomal instability (increased chromosome breakage) and defective DNA repair.

SWISS:
Q00597

Gene ID:
2176

	Database links: Entrez Gene: 2176Human Omim: 613899Human SwissProt: Q00597Human Unigene: 494529Human **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 Paraformaldehyde-fixed, paraffin embedded (human colon carcinoma); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (FANCC) Polyclonal Antibody, Unconjugated (SL13140R) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.