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Rabbit Anti-FAM123B antibody

SL6126R

Product Name:	FAM123B
Chinese Name:	肾母细胞瘤X蛋白抗体
Alias:	AMER1; FAM 123B; Family with sequence similarity 123B; FLJ39827; OSCS; Protein FAM123B; RP11 403E24.2; Wilms tumor gene on the X chromosome protein; Wilms tumor on the X; WTX; AMER1_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Pig,Cow,Horse,Rabbit,Sheep,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=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:	124kDa
Cellular localization:	The nucleuscytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human FAM123B/AMER1:281-380/1135
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:	The protein encoded by this gene upregulates trancriptional activation by the Wilms tumor protein and interacts with many other proteins, including CTNNB1, APC, AXIN1, and AXIN2. Defects in this gene are a cause of osteopathia striata with cranial sclerosis (OSCS).

Function:
Regulator of the canonical Wnt signaling pathway. Acts by specifically binding phosphatidylinositol 4,5-bisphosphate (PtdIns(4,5)P2), translocating to the cell membrane and interacting with key regulators of the canonical Wnt signaling pathway, such as components of the beta-catenin destruction complex. Acts both as a positive and negative regulator of the Wnt signaling pathway, depending on the context: acts as a positive regulator by promoting LRP6 phosphorylation. Also acts as a negative regulator by acting as a scaffold protein for the beta-catenin destruction complex and promoting stabilization of Axin at the cell membrane. Promotes CTNNB1 ubiquitination and degradation. Involved in kidney development.

Subunit:
Interacts with CTNNB1, AXIN1, LRP6, KEAP1, APC and BTRC. Interacts with SCF (SKP1-CUL1-F-box protein) E3 ubiquitin-protein ligase complexes containing BTRC and/or FBXW11. Identified in the beta-catenin destruction complex containing CTNNB1, APC, AXIN1 and AXIN2. Interacts with WT1.

Subcellular Location:
Cytoplasm. Cell membrane; Peripheral membrane protein; Cytoplasmic side. Nucleus. Note=Shuttles between nucleus and cytoplasm. Detected in nuclear paraspeckles that are found close to splicing speckles. Translocates to the cell membrane following binding to PtdIns(4,5)P2.

DISEASE:
Defects in FAM123B are the cause of osteopathia striata with cranial sclerosis (OSCS) [MIM:300373]. OSCS is an X-linked dominant sclerosing bone dysplasia that presents in females with macrocephaly, cleft palate, mild learning disabilities, sclerosis of the long bones and skull, and longitudinal striations visible on radiographs of the long bones, pelvis, and scapulae. In males this entity is usually associated with fetal or neonatal lethality.

Similarity:
Belongs to the Amer family.

SWISS:
Q5JTC6

Gene ID:
139285

Database links:

Entrez Gene: 139285Human

Entrez Gene: 72345Mouse

	Entrez Gene: 501584Rat Omim: 300647Human SwissProt: Q5JTC6Human SwissProt: Q7TS75Mouse **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.