

WTX Rabbit pAb

Catalog # AP99393

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q5JTC6
Other Accession	Q5JTC6
Host	Rabbit
Clonality	Polyclonal
Calculated MW	124029
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human WTX
Epitope Specificity	51-150/1135
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the Amer family.
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.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The protein encoded by this gene upregulates transcriptional 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).

Additional Information

Gene ID	139285
Other Names	FAM123B; OSCS; WTX; 2810002O09Rik; RGD1560322; AMER1_HUMAN; AMER1; Protein FAM123B; Wilms tumor gene on the X chromosome protein; AMER1_MOUSE; APC membrane recruitment protein 1; family with sequence similarity 123B; Wilms Tumor on the X; adenomatous polyposis coli membrane recruitment 1
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	AMER1
Synonyms	FAM123B, WTX
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.
Cellular 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
Tissue Location	Detected in fetal and adult kidney, brain and spleen.

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