abcam

Product datasheet

Recombinant Human Wilms Tumor Protein ab82233

製品の詳細

製品名	Recombinant Human Wilms Tumor Protein
精製度	> 90 % SDS-PAGE. Affinity purified
発現系	Baculovirus infected insect cells
アクセッション番号	NM_024426.4
タンパク質長	Full length protein
Animal free	No
由来	Recombinant
生物種	Human
配列	QDPASTCVPEPASQHTLRSGPGCLQQPEQQGVRDPGGIWAKL GAAEASAE RLQGRRSRGASGSEPQQMGSDVRDLNALLPAVPSLGGGGGCA LPVSGAAQ WAPVLDFAPPGASAYGSLGGPAPPPAPPPPPPPPPHSFIKQE PSWGGAEP HEEQCLSAFTVHFSGQFTGTAGACRYGPFGPPPPSQASSGQA RMFPNAPY LPSCLESQPAIRNQGYSTVTFDGTPSYGHTPSHHAAQFPNHS FKHEDPMG QQGSLGEQQYSVPPPVYGCHTPTDSCTGSQALLLRTPYSSDN LYQMTSQL ECMTWNQMNLGATLKGVAAGSSSSVKWTEGQSNHSTGYESDN HTTPILCG AQYRIHTHGVFRGIQDVRRVPGVAPTLVRSASETSEKRPFMC AYPGCNKR YFKLSHLQMHSRKHTGEKPYQCDFKDCERRFSRSDQLKRHQR RHTGVKPF QCKTCQRKFSRSDHLKTHTRTHTGKTSEKPFSCRWPSCQKKF ARSDELVR HHNMHQRNMTKLQLAL
予測される分子量	55 kDa
領域	1 to 517
タグ	DDDDK tag N-Terminus
配列の追加情報	Includes exon 5 and KTS. Flag-tagged isoform D.

特性

Our **Abpromise guarantee** covers the use of **ab82233** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

アプリケーション	SDS-PAGE EMSA Functional Studies Gel Supershift Assays
製品の状態	Liquid
備考	With the inclusion of exon 5, WT1 (KTS+) binds to both DNA and RNA and is RNase but not DNase sensitive (4). This form also co-localizes with splicing factors in a speckled nuclear particle, suggesting that the WT1 protein may function as both a transcription factor and a splicing regulator

前処理および保存

保存方法および安定性	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 7.9 Constituents: 0.75% Potassium chloride, 0.0154% DTT, 0.316% Tris HCl, 0.00584% EDTA, 20% Glycerol

関連情報

機能	Transcription factor that plays an important role in cellular development and cell survival. Regulates the expression of numerous target genes, including EPO. Plays an essential role for development of the urogenital system. Recognizes and binds to the DNA sequence 5'-CGCCCCCGC-3'. It has a tumor suppressor as well as an oncogenic role in tumor formation. Function may be isoform-specific: isoforms lacking the KTS motif may act as transcription factors. Isoforms containing the KTS motif may bind mRNA and play a role in mRNA metabolism or splicing. Isoform 1 has lower affinity for DNA, and can bind RNA.
組織特異性	Expressed in the kidney and a subset of hematopoietic cells.
関連疾患	Defects in WT1 are the cause of Frasier syndrome (FS) [MIM:136680]. FS is characterized by a slowly progressing nephropathy leading to renal failure in adolescence or early adulthood, male pseudohermaphroditism, and no Wilms tumor. As for histological findings of the kidneys, focal glomerular sclerosis is often observed. There is phenotypic overlap with Denys-Drash syndrome. Inheritance is autosomal dominant. Defects in WT1 are the cause of Wilms tumor 1 (WT1) [MIM:194070]. WT is an embryonal malignancy of the kidney that affects approximately 1 in 10'000 infants and young children. It occurs both in sporadic and hereditary forms. Defects in WT1 are the cause of Denys-Drash syndrome (DDS) [MIM:194080]. DDS is a typical nephropathy characterized by diffuse mesangial sclerosis, genital abnormalities, and/or Wilms tumor. There is phenotypic overlap with WAGR syndrome and Frasier syndrome. Inheritance is autosomal dominant, but most cases are sporadic. Defects in WT1 are the cause of nephrotic syndrome type 4 (NPHS4) [MIM:256370]. A renal disease characterized clinically by proteinuria, hypoalbuminemia, hyperlipidemia and edema. Kidney biopsies show non-specific histologic changes such as focal segmental glomerulosclerosis and diffuse mesangial proliferation. Some affected individuals have an

	inherited steroid-resistant form and progress to end-stage renal failure. Most patients with NPHS4 show diffuse mesangial sclerosis on renal biopsy, which is a pathologic entity characterized by mesangial matrix expansion with no mesangial hypercellularity, hypertrophy of the podocytes, vacuolized podocytes, thickened basement membranes, and diminished patency of the capillary lumen. Defects in WT1 are a cause of Meacham syndrome (MEACHS) [MIM:608978]. Meacham syndrome is a rare sporadically occurring multiple malformation syndrome characterized by male pseudohermaphroditism with abnormal internal female genitalia comprising a uterus and double or septate vagina, complex congenital heart defect and diaphragmatic abnormalities. Note=A chromosomal aberration involving WT1 may be a cause of desmoplastic small round cell tumor (DSRCT). Translocation t(11;22)(p13;q12) with EWSR1.
配列類似性	Belongs to the EGR C2H2-type zinc-finger protein family. Contains 4 C2H2-type zinc fingers.
細胞内局在	Nucleus. Cytoplasm. Shuttles between nucleus and cytoplasm; Nucleus > nucleoplasm and Nucleus speckle.

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