

## Product Information

### Anti-WTX (N-terminal)

produced in rabbit, affinity isolated antibody

Product Number **SAB4200184**

### Product Description

Anti-WTX (N-terminal) is produced in rabbit using as the immunogen a synthetic peptide corresponding to a sequence at the N-terminal of human WTX (GeneID: 139285), conjugated to KLH. The corresponding sequence is highly conserved (77% sequence identity) in mouse and rat WTX. The antibody is affinity-purified using the immunizing peptide immobilized on agarose.

Anti-WTX (N-terminal) specifically recognizes human WTX. The antibody may be used in several immunochemical techniques including immunoblotting (~190 kDa). An additional band at ~150 kDa may be observed corresponding to an alternatively spliced WTX form. Detection of the WTX band by immunoblotting is specifically inhibited by the WTX immunizing peptide.

Wilms tumor, the most common pediatric kidney cancer, is a pluripotent tumor that arises from a kidney-specific stem cell population.<sup>1</sup> Loss of function mutations in *WT1*, a transcription factor with multiple roles in the developing kidney, are present in 5-10% of cases, often in association with activating mutations in  $\beta$ -catenin, a key regulator of WNT signaling. *WTX* (also known as FAM123B) is a recently identified X chromosome gene that is targeted by deletions and truncations in up to 30% of Wilms tumors.<sup>2,3</sup> *WTX* gene encodes a 190 kDa protein and a 150 kDa form resulting from alternative splicing at the N-terminus. All known *WTX* mutations are somatic, targeting the single X chromosome in males or the active X chromosome in females, thus leading to its complete inactivation. *WTX* has been shown to form a complex with  $\beta$ -catenin, AXIN1,  $\beta$ -TrCP2, and APC. In cultured cells, *WTX* negatively regulates the WNT/ $\beta$ -catenin signaling by promoting  $\beta$ -catenin ubiquitination and thus targets its degradation by the proteasome.<sup>4</sup> Loss of *WTX* function in Wilms tumor results in stabilization of  $\beta$ -catenin and its translocation to the nucleus. *WTX* shuttles between the cytoplasm and nucleus and is present in a distinct subnuclear compartment implicated in transcription and RNA processing.<sup>5</sup> *WTX* co-localizes with p54nrb/NONO and binds *WT1* to modulate its activity, suggesting that *WTX* may play a role in the transcriptional regulation of cell differentiation.

### Reagent

Supplied as a solution in 0.01 M phosphate buffered saline, pH 7.4, containing 15 mM sodium azide.

Antibody concentration: ~1.0 mg/mL

### Precautions and Disclaimer

For R&D use only. Not for drug, household, or other uses. Please consult the Safety Data Sheet for information regarding hazards and safe handling practices.

### Storage/Stability

For continuous use, store at 2–8 °C for up to one month. For extended storage, freeze in working aliquots. Repeated freezing and thawing, or storage in “frost-free” freezers, is not recommended. If slight turbidity occurs upon prolonged storage, clarify the solution by centrifugation before use. Working dilutions should be discarded if not used within 12 hours.

### Product Profile

**Immunoblotting:** a working concentration of 1-2  $\mu$ g/mL is recommended using HEK-293T cell lysates overexpressing human *WTX*.

**Note:** In order to obtain best results in various techniques and preparations, it is recommended to determine optimal working dilutions by titration.

### References

1. Rivera, M.N., and Haber, D.A., *Nat. Rev. Cancer Res.*, **5**, 699-712 (2005).
2. Rivera, M.N., et al., *Science*, **315**, 642-645 (2007).
3. Han, M., et al., *Genes Chromosomes Cancer*, **46**, 906-913 (2007).
4. Major, M.B., et al., *Science*, **316**, 1043-1046 (2007).
5. Rivera, M.N., et al., *Proc. Natl. Acad. Sci. USA*, **106**, 8338-8343 (2008).

VS,ER,KAA,PHC,MAM 07/19-1