

Gene Section

Short Communication

SET (SET translocation (myeloid leukemia-associated))

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Identity

Other names: I2PP2A, I-2PP2A (Phosphatase 2A inhibitor); TAF-IBETA (Template activating factor I) PHAPII (HLA-DR associated protein II); IGAAD (Inhibitor of granzyme A-activated Dnase)

HGNC (Hugo): SET

Location: 9q34

Local order: from centromere to telomere: SET, ABL1, NUP214 (alias CAN), NOTCH1 (alias TAN1).

DNA/RNA

Description

SET encompasses 6.81 kb on the genomic DNA; 8 exons.

Transcription

2577 bp mRNA.

Protein

Description

Two isoforms; isoform 1 (TAF1 alpha) 290 amino acids, 33.5 kDa; isoform 2 (TAF1 beta) - 277 amino acids, 23.1 kDa.

Expression

Widely expressed; highly expressed in Wilms' tumor.

Localisation

Cytoplasmic and nuclear; in the cytoplasm, found both in the cytosol and associated with the endoplasmic reticulum.

Function

Multitasking protein, involved in apoptosis, transcription, nucleosome assembly, and histone binding; potent inhibitor of protein phosphatase 2A; inhibits also EP300/CREBBP and PCAF-mediated acetylation of histones (HAT) - predominantly H4 - and nucleosomes; HAT inhibition leads to silencing of HAT-dependent transcription and prevents active demethylation of DNA.

Homology

Belongs to the nucleosome assembly protein (NAP) family.

Implicated in

t(9;9)(q34;q34) --> SET-NUP214 (alias CAN)

Disease

Acute undifferentiated leukemia (AUL); only one case described so far.

Cytogenetics

Normal karyotype; may be overlooked.

Hybrid/Mutated gene

5' SET - 3' NUP214.

Abnormal protein

The SET-NUP214 (alias CAN) fusion protein consists of almost the whole SET protein fused to the C-terminus of NUP214.

Oncogenesis

SET-NUP214 leads to disorganization of nuclear export.

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