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Gene Section
Mini Review

NR4A3 (nuclear receptor subfamily 4, group A, member 3)

Göran Stenman

Lundberg Laboratory for Cancer Research, Department of Pathology, G&ouml (GS)

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Identity

Other names: TEC; NR4A3; NOR1; MINOR; CSMF; CHN

HGNC (Hugo): NR4A3

Location: 9q22

DNA/RNA

Description

Spans about 46 kb; 9 exons.

Transcription

Four transcript variants (6382 bp, 5635 bp, 4983bp and 2588 bp).

Protein

Description

Three isoforms (637 aa, 626 aa and 443 aa); N-terminal activation domain; central bipartite zinc finger DNA-binding domain; C-terminal ligand binding domain.

Expression

Most tissues, highest expression levels in the central nervous system.

Localisation

Nuclear.

Function

Orphan nuclear receptor; implicated in the control of cell proliferation as an immediate-early protein and in differentiation and apoptosis; binds as a monomer to the DNA-response element NBRE; activate transcription constitutively when bound to NBRE; the homeotic protein Six3 is a coactivator of TEC.

Homology

Belongs to the steroid/thyroid receptor gene super family; highly homologous to the nuclear receptors NGFI-B and NURR1.

Implicated in

Extraskeletal myxoid chondrosarcoma with t(9;22)(q22;q12)

Disease

Rare type of sarcoma (2.3% of all soft tissue sarcomas) characteristically involving the deep, soft tissues of the extremities; morphological resemb-lance to embryonic cartilage.

Hybrid/Mutated gene

EWSR1-TEC

Abnormal protein

N-terminal transactivation domain of EWSR1 fused to the entire TEC protein.

Extraskeletal myxoid chondrosarcoma with t(9;17)(q22;q11)

Disease

Rare type of sarcoma (2.3% of all soft tissue sarcomas) characteristically involving the deep, soft tissues of the extremities; morphological resemb-lance to embryonic cartilage.

Hybrid/Mutated gene

TAF2N-TEC

Abnormal protein
N-terminal transactivation domain of TAF2N fused to the entire TEC protein.

Extraskeletal myxoid chondrosarcoma with t(9;15)(q22;q21)

Disease
Rare type of sarcoma (2.3% of all soft tissue sarcomas) characteristically involving the deep, soft tissues of the extremities; morphological resemb-lance to embryonic cartilage.

Hybrid/Mutated gene
TCF12-TEC

Abnormal protein
N-terminal domain of TCF12 fused to the entire TEC protein; the N-terminal of TCF12 shows no sequence homology to the N-terminals of EWSR1 or TAF2N.

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