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Gene Section
Mini Review

TLX3 (T-cell leukemia, homeobox protein 3)

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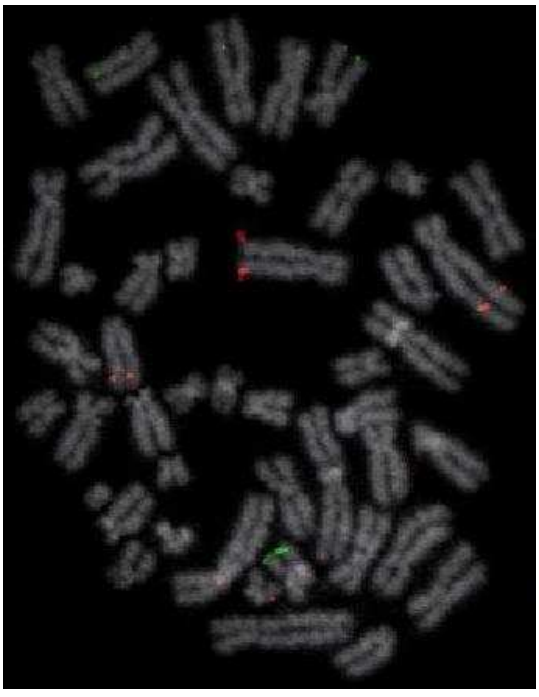
Identity

Other names: RNX; HOX11L2

HGNC (Hugo): TLX3

Location: 5q35.1



TLX3 (5q35.1): FISH with BAC clone ctb-45L16 (red) showing a breakpoint signal in the consensus breakpoint region split between der(5) and der(14) partners. Analysis was performed on the pediatric T-ALL cell line HPB-ALL (DSMZ ACC-483) which carries t(5;14)(q35.1;q32.2) as part of a complex 4-way rearrangement involving chromosomes 1 and 16. Normally (in the absence of secondary translocations) the breakpoints on both partners lie closely equidistant to the q-arm telomeres rendering the translocation cryptic and difficult to detect even with chromosome painting.

DNA/RNA

Description

3 exons on 2.23 kb.

Transcription

In a centromeric --> telomeric orientation; 876 bp mRNA (coding).

Protein

Description

291 amino acids, 31.8 kDa; contains one homeobox domain (residues 166-221).

Expression

Narrowly restricted to brain.

Localisation

Probably nuclear.

Function

Murine Tlx-3 and Tlx-1 together sustain expression of Drg-11, and control development of somatic and visceral relay sensory neurons.

Homology

With homeobox genes, especially with those of the NK-like family.

Implicated in

t(5;14)(q35;q32) in T-ALL-->TLX3 - BCL11B

Disease
T-cell acute lymphocytic leukemia (T-ALL).

Prognosis
TLX3 expression may denote poor prognosis.

Cytogenetics
Cryptic translocation detectable by locus specific FISH. t(5;14) may exclude del(1)(p32) SIL-TAL1 fusion.

Hybrid/Mutated gene
5' TLX3-3' BCL11B on der(14).

Oncogenesis
Ectopic expression in T-cells

t(5;14)(q35;q11) in T-ALL --> TLX3 – TCRD.

Disease
T-cell acute lymphocytic leukemia (T-ALL).

Cytogenetics
Apparent variant of t(5;14).

Hybrid/Mutated gene
5' TLX3-TCRD on der(14)

Breakpoints

Within the upstream region or within the neighboring gene RanBP17.

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