TLX3 (T-cell leukemia, homeobox protein 3)

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Identity

Other names: RNX; HOX11L2
HGNC (Hugo): TLX3
Location: 5q35.1

DNA/RNA

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.

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.
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

\textbf{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

\begin{itemize}
\item RanBP17
\item TLX3
\end{itemize}

Within the upstream region or within the neighboring gene RanBP17.

References


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