

Atlas of Genetics and Cytogenetics in Oncology and Haematology

t(7;9)(q34;q32)

Clinics and Pathology

Disease	Specifically associated with T-cell Acute lymphoblastic leukemia (T-ALL)
Epidemiology	The t(7;9)(q34;q32) is present in two cases of a serie of 5 patients with 7q34 involvment.
Clinics	Children with large mediastinal mass
Cytology	The hematological feature of the T-ALL with rearrangement of 7q34 shows high WBC (range 95 to 400 x 10 ⁹ /L)

Cytogenetics

Cytogenetics	9q32 is a partner of 7q34. The other partners are 1p34,1p32,
Morphological	9q34,10q24,11p13, 15q22 and 19p13
Additional anomalies	del(6)(q21), del(4)(q31q35)

Genes involved and Proteins

Gene Name [TCRB \(T-cell receptor beta-chain\)](#)

Location	7q35
Dna / Rna	The TRB locus at 7q35 spans 685 Kb The locus contains 2 types of coding elements : TCR elements (64-67 variable genes TRBV, 2 clusters of diversity, joining and constant segments) and 8 trypsinogen genes. A portion of the TCRB locus has been duplicated and translocated to the chromosome 9 at 9p21.
Protein	T cell receptor beta chains.

Gene Name [TAL2](#)

Location	9q32
Dna / Rna	The TAL2 gene is located at q32
Protein	TAL2 potentially encodes a basic helix-loop-helix (bHLH) phosphoprotein (size 108 amino acids) that is highly related to those specified by TAL1 and LYL1 also implicated in T-ALL. The bHLH protein interacts with the product of RBTN1 and RBTN2 (cysteine-rich LIM motifs)

Result of the chromosomal anomaly

Hybrid gene	TAL2 is transcriptionally activated by t(7;9)(q34;q32) in T-ALL.The chromosome 9 breakpoints of the t(7;9)(q34;q32) occur 33 kbp downstream of sequences that encode the TAL2 HLH domain.
Description	Translocated TAL2 are juxtaposed with transcriptional regulatory elements within the T-cell receptor beta-chain locus.

Fusion Protein Note No fusion protein.

Oncogenesis The TAL2 transcription is activated ectopically in lymphoid cells and the

inappropriate expression of TAL2 in these cells promotes development of T-ALL. Normally, the TAL genes are not expressed in the thymus. The TAL genes become activated and expressed in the thymus upon chromosomal translocation which ultimately leads to the development of T-ALL.

The (7;9) translocation express a TAL2 gene product of 108 amino acids. In leukemic cells this product exists in both a phosphorylated and an unphosphorylated form. Serine residue 100 is the major site of TAL2 phosphorylation in vivo. And it serves as an effective in vitro substrate for MAP kinases such as ERK1.

TAL2 polypeptides interact in vivo with the [E2A](#) gene products to form HLH heterodimers that bind DNA, the result is the E2A inactivation. The E2A products are transcriptional factors implicated in the B and T cell development.

TAL2 product was also shown to bind with a GTP binding protein (DRG). The properties of TAL2 broadly resemble those described previously for TAL1 and therefore support the idea that both encoded proteins promote T-ALL by a common mechanism and the malignant potential of these proteins is likely to reside within their HLH domains though the inactivation of E2A.

External links

Other database [t\(7;9\)\(q34;q32\)](#) [Mitelman database \(CGAP - NCBI\)](#)
 Other database [t\(7;9\)\(q34;q32\)](#) [CancerChromosomes \(NCBI\)](#)

To be noted

Additional cases are needed to delineate the epidemiology of this rare entity:
you are welcome to submit a paper to our new [Case Report](#) section.

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