

## t(11;14)(p15;q11)

### Clinics and Pathology

<b>Disease</b>	T-cell Acute lymphoblastic leukemia T-ALL
Phenotype / cell stem origin	Apparently, this translocation is not restricted to a single maturational stage. Among five cases reported in the literature: 2 of 4 cases tested coexpressed CD4 and CD8 antigens and 2 cases showed a more immature phenotype like Pro T-ALL: cytCD3+ CD3- CD4- CD8- CD34+
Epidemiology	The t(11 ;14)(p15 ;q11) occurs in less than 1% of patients with T-cell ALL
Cytology	There is limited knowledge about the clinical and biologic characteristics of patients in whom this translocation is present, all of them have childhood T-cell ALL In contrast to cases with the t(11 ;14)(p13 ;q11) only one patient had a markedly elevated leucocyte count at diagnosis.
Prognosis	Remarkably, no patient with this translocation has sustained durable remission.

### Cytogenetics

Cytogenetics	This translocation is not to be confused with the <a href="#">t(11 ;14)(p13 ;q11)</a> .
Morphological	
Additional anomalies	del(6q), i(17)(q10)

### Genes involved and Proteins

<b>Gene Name</b>	Rhombotin-1 (= LMO1 = LIM domain only 1, TTG1, RBTN1)
Location	11p15 1214 bp. This gene belongs to the Rhombotin family: RBTN1, <a href="#">RBTN2</a> , RBTN3. Complete characterisation of these genes in man and mouse shows that all three encode cysteine-rich proteins with typical LIM domains. The exon organisation of RBTN1 and RBTN3 are similar, both having an intron, absent from the RBTN2 gene, in the LIM2 encoding region.
Dna / Rna	
Protein	RBTN1 and RBTN3 derived proteins have 98% identity in the LIM domain; LMO1 derived protein is a 46kd nuclear protein. Comparison of the sequence of the human and mouse protein LMO1 shows that the main conserved sequence is a tandemly duplicated cystein-rich-region called LIM domain. LIM domain might facilitate protein-protein interaction which modulate transcription via intermolecular competitive binding between LIM domain and certain DNA-binding-transcription factors. The LDB1/NLI is a phosphoprotein and binds to LMO1 in its phosphorylated state and essentially all the LMO1 and LDB1 protein in the T cell is part of the complex. The stem cell leukemia ( <a href="#">SCL</a> ) transcription factor is also a partner for LMO1 and LMO2 proteins. RBTN1 and RBTN3 proteins have the same expression pattern in mouse development, since

both genes show high expression in the brain, but little lymphoid expression. RBTN2 expression is more ubiquitous.

**Gene Name** [TCR -alpha](#) and [TCR-delta](#)

**Location** 14q11.2

The size of TCR alpha/delta locus is about 1 Mb. The TCR delta variable (V) diversity (D) joining (J) and constant region genes are situated within the TCR alpha locus between the TCR alpha V and the TCR alpha J segments. The TCR delta locus contains three D segments and four J segments, whereas the TCR alpha J regions spans approximately 80 Kb and contains at least 61 segments. The TCR alpha/delta locus is transcribed in a centromer to telomer direction.

**Protein** T-cell receptor

### Result of the chromosomal anomaly

**Hybrid gene** Description The translocation t(11 ;14)(p15 ;q11) occurs at a T-cell receptor joining J delta segment, 12 kb upstream of the constant C delta gene and 98 kb upstream of the C alpha gene at chromosome band 11q11. Rhombotin gene contains oligomers that span the breakpoint region. This breakpoint region possesses a consensus heptamer : it appers to function as a 23-signal and to synapse with a 12-signal on chromosome 14 in the translocation t(11 ;14)(p15 ;q11) but the recombinaison frequency is 20-fold lower than LMO2 and about 530-fold lower than for a normal 23-signal. The 23-signal is a part, as well as the 12-signal, of the RAG complex whose normal function is to initiate the breaks of V(D)J recombinaison.

### Fusion

**Protein** No fusion protein.

Description

**Oncogenesis** Erroneous V(D)J joining process and alteration of the transcription networks.

### External links

Other database [t\(11;14\)\(p15;q11\)](#) [Mitelman database \(CGAP - NCBI\)](#)

Other database [t\(11;14\)\(p15;q11\)](#) [CancerChromosomes \(NCBI\)](#)

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