

# Leukaemia Section

## Mini Review

### t(11;14)(q13;q32)

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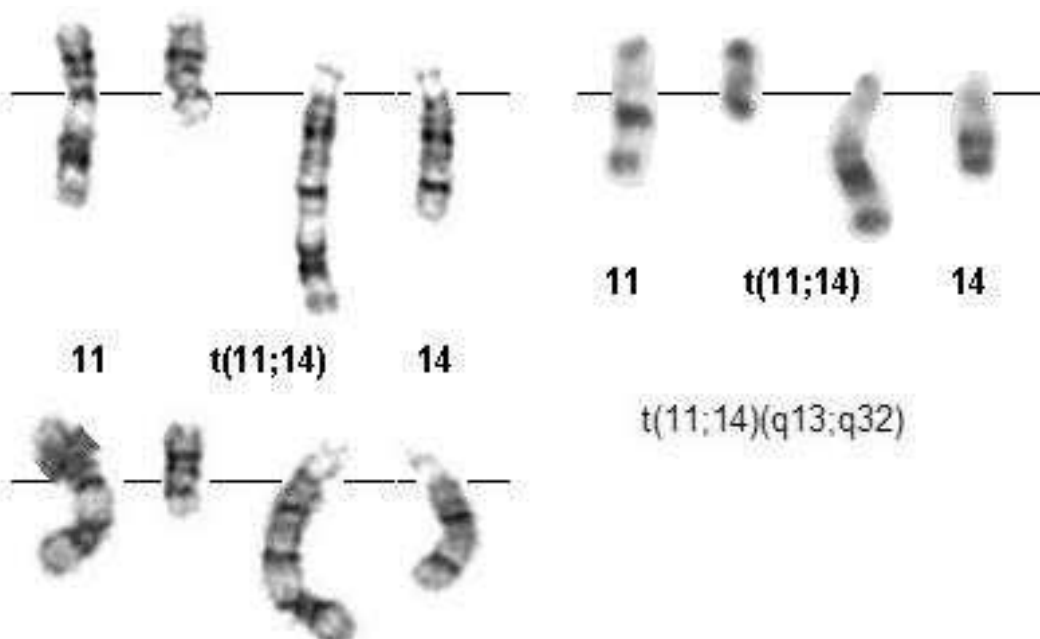
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## Identity



t(11;14)(q13;q32) G-banding (left) - Courtesy Diane H. Norback, Eric B. Johnson, Sara Morrison-Delap Cytogenetics at the Waisman Center and R-banding (right) - Editor.

**Note:** t(11;14) is mainly found in mantle cell lymphoma, but also in B-prolymphocytic leukaemia, in plasma cell leukaemia, in splenic lymphoma with villous lymphocytes, in chronic lymphocytic leukaemia, and in multiple myeloma, herein briefly described; all these diseases involve a B-lineage lymphocyte

## Clinics and pathology

### Disease

Mantle cell lymphoma.

### Phenotype / cell stem origin

B-cell non Hodgkin lymphoma of the low to intermediate grade.

### Epidemiology

Annual incidence  $5/10^6$ ; median age: 65 yrs.

### Clinics

Advanced disease.

### Prognosis

Median survival: 3 to 4 yrs.

**Disease**

B-prolymphocytic leukaemia.

**Phenotype / cell stem origin**

Chronic lymphoproliferative disorder affecting mature B-cells.

**Epidemiology**

Rare disease; median age 70 yrs.

**Clinics**

Patients often present with advanced stage disease.

**Prognosis**

Median survival: 3 yrs.

**Disease**

Plasma cell leukaemia.

**Phenotype / cell stem origin**

Proliferation involving plasma cells.

**Epidemiology**

Rare disorder.

**Prognosis**

Median survival is less than a yr.

**Disease**

Splenic lymphoma with villous lymphocytes.

**Phenotype / cell stem origin**

Chronic B-cell lymphoproliferation.

**Epidemiology**

Rare disorder; median age: 70 yrs.

**Clinics**

Relatively benign clinical course.

**Prognosis**

80% 5-yr survival.

**Disease**

Chronic lymphocytic leukaemia.

**Phenotype / cell stem origin**

Chronic B-cell lymphoproliferation.

**Epidemiology**

Annual incidence 30/10<sup>6</sup>; median age: 60-80 yrs.

**Clinics**

Often a slow evolutive disease.

**Prognosis**

Highly variable according to the staging: from staging A: survival not reduced compared to age matched population, to staging C: median survival of 2 yrs.

**Disease**

Multiple myeloma.

**Phenotype / cell stem origin**

Malignant plasma cell proliferation (terminally differentiated B-cell).

**Epidemiology**

Annual incidence: 30/10<sup>6</sup>; median age: 60 yrs.

**Prognosis**

Median survival: 3 yrs.

**Cytogenetics****Cytogenetics, morphological**

t(11;14) has earlier been thought to be the hallmark of the mantle cell lymphoma; actually, the frequency of t(11;14) is: 50-70% in mantle cell lymphoma, 10-20% in B-prolymphocytic leukaemia, in plasma cell leukaemia, and in splenic lymphoma with villous lymphocytes, and 2-5% in chronic lymphocytic leukaemia, and in multiple myeloma.

**Cytogenetics, molecular**

In particular interphase cytogenetics, are relevant in these diseases with an usually low mitotic index.

**Additional anomalies**

Sole anomaly in only 10% of cases; part of a complex karyotype in 2/3 of cases; numerous recurrent anomalies found conjointly (which is the primary?), particularly: +3, +7, del(9p), +18, +mar, found in about 10% of cases each; other: del(1p), del(6q), del(7q), -8, +12, del(13q), del(17p).

**Variants**

Three way complex t(11;14;Var) exist and showed that the crucial event lies on der(14).

**Genes involved and Proteins****BCL1**

**Location:** 11q13

**DNA / RNA**

5 exons.

**Protein**

Encodes the cyclin D1; role in the cell cycle control: G1 progression and G1/S transition.

**IgH**

**Location:** 14q32

**Results of the chromosomal anomaly****Hybrid gene****Description**

5' BCL1 translocated on chromosome 14 near JH (junctions genes of IgH) and C in 3'; the breakpoint in BCL1 is in MTC (major translocation cluster), centromeric to the gene (in 5'), in 80% of cases, or dispersed in mTC1, 2, or 3 in 5' of the gene or in the 3' untranslated region of exon 5.

## **Fusion protein**

### **Description**

No fusion protein, but promoter exchange; the immunoglobulin gene enhancer stimulates the expression of BCL1.

### **Oncogenesis**

Overexpression of BCL1 accelerates passage through the G1 phase.

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