

# Gene Section

## Mini Review

# ETV6 (ETS variant gene 6 (TEL oncogene))

**Serge Pierrick Romana**

Service de Cytogenetique (Unite de Cytogenetique Moleculaire), Hopital Necker-Enfants-Malades, 149, rue de Sevres, 75015 Paris, France (SPR)

Published in Atlas Database: December 1999

Online updated version : <http://AtlasGeneticsOncology.org/Genes/ETV6ID38.html>  
DOI: 10.4267/2042/37554This work is licensed under a Creative Commons Attribution-Noncommercial-No Derivative Works 2.0 France Licence.  
© 1999 *Atlas of Genetics and Cytogenetics in Oncology and Haematology*

## Identity

**Other names:** TEL (translocation ets leukemia)**Location:** 12p13.1

## DNA/RNA

### Description

The gene spans a region of 240 kb.

### Transcription

Transcription is from telomere to centromere; there are three species of transcripts: 2400 kb, 4300 kb and 6200 kb; the gene encodes for a 1356 kb cDNA.

## Protein

### Description

Two TEL human protein isoforms have been characterized: one of 53 kDa and one of 57 kDa; these correspond respectively to translational initiation from the second in frame methionine (codon 43) and from the first in frame methionine (codon 1); it has been demonstrated that these two isoforms are phosphorylated; these proteins belong to the ETS transcription factors family characterized by the presence of 85 amino acids, the ETS domain; this domain is responsible for the sequence specific DNA-binding activity GGAA/T flanked by a 5-8 nucleotides contributing to the specificity of each proteins ETS members; TEL possesses an N-terminal domain called NH<sub>2</sub> terminal conserved region (NCR) which is found in other ETS proteins. This TEL domain unlike most of the other NCR domains is responsible for the TEL protein homotypic oligomerization capacity.

## Expression

In mouse, the TEL proteins are more expressed in the neural tube, in cranial node, in mesenchymateus tissue adjacent to the primitive intestine.

## Localisation

Immunofluorescent experiences revealed a nucleus localization of the TEL proteins.

## Function

TEL proteins belong to the ETS family transcription factors; different mouse KO experiences have demonstrated that TEL are important in the vitelline angiogenesis and in the bone marrow hematopoiesis.

## Implicated in

### Leukemia and sarcoma

## References

Wessels JW, Fibbe WE, van der Keur D, Landegent JE, van der Plas DC, den Ottolander GJ, Rozendaal KJ, Beverstock GC. t(5;12)(q31;p12). A clinical entity with features of both myeloid leukemia and chronic myelomonocytic leukemia. *Cancer Genet Cytoogenet.* 1993 Jan;65(1):7-11

Golub TR, Barker GF, Bohlander SK, Hiebert SW, Ward DC, Bray-Ward P, Morgan E, Raimondi SC, Rowley JD, Gilliland DG. Fusion of the TEL gene on 12p13 to the AML1 gene on 21q22 in acute lymphoblastic leukemia. *Proc Natl Acad Sci U S A.* 1995 May 23;92(11):4917-21

Peeters P, Raynaud SD, Cools J, Wlodarska I, Grosgeorge J, Philip P, Monpoux F, Van Rompaey L, Baens M, Van den Berghe H, Marynen P. Fusion of TEL, the ETS-variant gene 6 (ETV6), to the receptor-associated kinase JAK2 as a result of t(9;12) in a lymphoid and t(9;15;12) in a myeloid leukemia. *Blood.* 1997 Oct 1;90(7):2535-40

Peeters P, Wlodarska I, Baens M, Crikel A, Selleslag D, Hagemeijer A, Van den Berghe H, Marynen P. Fusion of ETV6 to MDS1/EVI1 as a result of t(3;12)(q26;p13) in myeloproliferative disorders. *Cancer Res.* 1997 Feb 15;57(4):564-9

Suto Y, Sato Y, Smith SD, Rowley JD, Bohlander SK. A t(6;12)(q23;p13) results in the fusion of ETV6 to a novel gene, STL, in a B-cell ALL cell line. *Genes Chromosomes Cancer.* 1997 Apr;18(4):254-68

Chase A, Reiter A, Burci L, Cazzaniga G, Biondi A, Pickard J, Roberts IA, Goldman JM, Cross NC. Fusion of ETV6 to the caudal-related homeobox gene CDX2 in acute myeloid leukemia with the t(12;13)(p13;q12). *Blood.* 1999 Feb 1;93(3):1025-31

Cools J, Bilhou-Nabera C, Wlodarska I, Cabrol C, Talmant P, Bernard P, Hagemeijer A, Marynen P. Fusion of a novel gene, BTL, to ETV6 in acute myeloid leukemias with a t(4;12)(q11-q12;p13). *Blood.* 1999 Sep 1;94(5):1820-4

Yagasaki F, Jinnai I, Yoshida S, Yokoyama Y, Matsuda A, Kusumoto S, Kobayashi H, Terasaki H, Ohyashiki K, Asou N, Murohashi I, Bessho M, Hirashima K. Fusion of TEL/ETV6 to a novel ACS2 in myelodysplastic syndrome and acute myelogenous leukemia with t(5;12)(q31;p13). *Genes Chromosomes Cancer.* 1999 Nov;26(3):192-202

---

*This article should be referenced as such:*

Romana SP. ETV6 (ETS variant gene 6 (TEL oncogene)). *Atlas Genet Cytogenet Oncol Haematol.* 1999; 3(4):181-182.

---