Atlas of Genetics and Cytogenetics in Oncology and Haematology

OPEN ACCESS JOURNAL AT INIST-CNRS

# Gene Section
## Mini Review

# LAZ3 (Lymphoma Associated Zinc finger on chromosome 3)

**Jean-Pierre Kerckaert**

Unite 124 INSERM, Institut de Recherche sur le Cancer, Place de Verdun, 59045 Lille cedex, France (JPK)

Published in Atlas Database: September 1998

Online updated version : http://AtlasGeneticsOncology.org/Genes/BCL6ID20.html
DOI: 10.4267/2042/37472

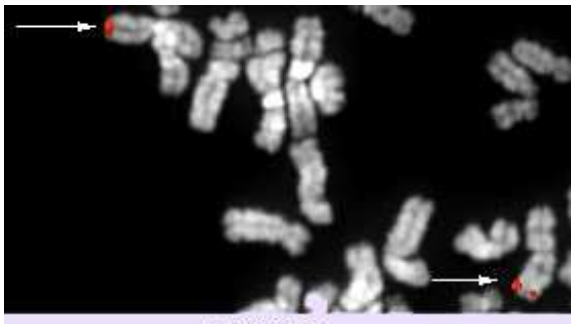


## Identity

**Other names:** BCL6 (B-Cell Lymphoma 6)

**Location:** 3q27

**Local order:** Gene orientation: telomere - 5' LAZ3 3' - centromere.




BCL6 (3q27) - Courtesy Mariano Rocchi.

## DNA/RNA

### Description

Spans on a 25 kb genomic fragment. 11 exons; the two first exons 1A and 1B are alternative; translational ATG in exon 3.

### Transcription

3.8 kb mRNA.

## Protein

### Description

706 amino acids; 79 kDa; BTB/POZ Zinc finger protein: N-term BTB/POZ domain (130-aa protein-protein interaction motif and repressing domain) and C-term with 6 Krüppel-like zinc fingers (sequence specific DNA binding domain).

### Expression

Normally expressed in germinal center B and T cells, in skeletal muscle cells and in keratinocytes.

### Localisation

Nuclear dots.

### Function

Sequence-specific DNA binding transcriptional repressor; consensus DNA-binding site: TTC(C/T)T(A/C)GAA; the LAZ3/BCL6 protein mediates transcriptional repression by recruiting (through the BTB/POZ domain) a nuclear hormone receptor co-repressor (SMRT) and histone deacetylation; it is required for the formation of germinal centers and the Th2 mediated response.

### Homology

BTB/POZ - Zinc Finger proteins (PLZF, HIC1, KUP, BAZF, ttk (drosophila); BrC (drosophila)...).

## Implicated in

### 3q27 rearrangements / NHL (non Hodgkin lymphomas)

**Disease**

NHL: in 30-40% of diffuse large cell lymphoma, 5-14% of follicular lymphoma.

**Prognosis**

Still controversial (favourable in BCL6- vs BCL2-rearranged non Hodgkin lymphomas according to some studies or not significative in other reports).

**Cytogenetics**

3q27 rearrangements are diverse : translocations, micro-deletions, point mutations and hypermutation); about half of 3q27 translocations Ig genes at 14q32 (IgH), 2p12 (IgK) and 22q12 (IgL) (e.g. t(3;14)(q27;q32)); the other half includes a variety of other chromosomal regions (1q21, 2q21, 4p11, 5q31, 6p21, 7p12, 8q24, 9p13, 11q13, 11q23, 12q11, 13q14-21, 14q11, 15q21, 16p11...); frequent bi-allelic alterations (translocation and deletion or mutation on the non-translocated allele).

**Hybrid/Mutated gene**

Hybrid gene and transcripts are formed following promoter substitution between LAZ3 and its different partner: Ig-LAZ3 in t(3;14)(q27;q32); RHOH-LAZ3 in t(3;4)(q27;p13); Histone H4-LAZ3 in t(3;6)(q27;p21); OBF1-LAZ3 in t(3;11)(q27;q23), L-Plastin-LAZ3 in t(3;13)(q27;q14-21); chimeric transcripts are generally detected containing the 5' part of the gene partner fused to the normal LAZ3 exon 2 splice acceptor site; in some cases reciprocal chimeric transcripts driven by the 5' regulatory region of LAZ3 fused to the partner gene coding region, have been characterised.

**Abnormal protein**

No fusion protein.

# Breakpoints

**Note**

Clustered in a 3,3 kb EcoRI fragment (MTC) includind exon 1A and intron 1.

# References

Kerckaert JP, Deweindt C, Tilly H, Quief S, Lecocq G, Bastard C. LAZ3, a novel zinc-finger encoding gene, is disrupted by recurring chromosome 3q27 translocations in human lymphomas. Nat Genet. 1993 Sep;5(1):66-70

*This article should be referenced as such:*

Kerckaert JP. LAZ3 (Lymphoma Associated Zinc finger on chromosome 3). Atlas Genet Cytogenet Oncol Haematol. 1999; 3(1):4-5.